THE ESOPHAGUS
Medical and Surgical Management

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MEDICAL AND SURGICAL MANAGEMENT

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This book is dedicated to

Patricia Hatfield Benedict, M D

Sally Cantwell Nardi

and

Antenisca R Nardi

whose patience and encouragement have made it possible

FOREWORD

Systematized knowledge about the esophagus has grown at a leisurely pace partly because the organ itself is so inaccessible and partly because experience with clinical cases has been dispersed among surgeons, physicians endoscopists and roentgenologists Many of the previous monographs on the esophagus have had a particularized slant because very few doctors of any ilk have looked at the whole esophagus in health and disease This undertaking by Dr Benedict and Dr Nardi is designed to provide such a unified approach

Writing as experts the authors naturally voice impatience with what they consider complacency on the part of the physician when he is confronted by a patient who is having difficulty in swallowing From an over all point of view however one can only be encouraged by such tangible evidence of the development of helpful knowledge and useful technical undertakings which has taken place in two decades Much of this development has been a product of the steadily increasing scope of thoracic surgery Open thoracotomy has permitted a comfortable and studied appraisal of lesions of the esophagus in the living patient and so changed esophageal surgery from a blind and crude undertaking to an orderly and productive application of standardized technics

There is still much to be learned about the esophagus and its lesions A monographic presentation of the existing knowledge and experience should provoke as many questions as it can provide satisfactory answers In this important aspect the authors have followed the only tolerable course — humility in the face of the unknown

EDWARD D CHURCHILL, M D

Preface

This book is planned to give the reader a reasonably complete but concise knowledge of the diagnosis and treatment of esophageal disease. Until recently the esophagus was a rather neglected organ because almost nothing could be done about it surgically. Now, however, it is frequently approached with confidence by the *thoracic surgeon* and partly under his stimulation x-ray technics and endoscopic methods have been improved. Therefore the time seems appropriate to write up our experiences as practiced at the Massachusetts General Hospital and as taught at the Harvard Medical School.

We wish to acknowledge with thanks the untiring secretarial help of Miss Priscilla Boujoukos and Miss Ann Richer, and the excellent illustrative work of Mrs. Edith Tagrin.

Some of the illustrations, including the esophagoscopic views shown in color in Plates VI and IX, first appeared in *Endoscopy* by Edward B. Benedict, M.D., The Williams and Wilkins Company, 1951. We are deeply grateful to Williams and Wilkins for their courtesy in making these illustrations available to us.

The authors appreciate the constant co-operation of the publisher's staff.

Edward B. Benedict, M.D.
George L. Nardi, M.D.

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INTRODUCTION

I

When one considers how important eating is one wonders why so little attention has been paid to the organ by which food is conveyed into the stomach Even if we don t live to eat as many people seem to we must eat to live hence the great importance of the esophagus We hope this book will encourage more doctors to give increased consideration to the esophagus Too often patients with persistent dysphagia are diagnosed as hysterical The precaution of X-ray examination is also overlooked

All physicians and many laymen have heard of cancer of the esophagus Advances in thoracic surgery are such that the disease is no longer hopeless There is no longer any excuse not to attempt treatment Curative resections for carcinoma of the esophagus are now a fact and palliative resections have proved worth while in many cases

Moreover many other diseases of the esophagus may be diagnosed by X ray and by esophagoscopy and may be treated by general or specific medical methods — X ray therapy in inoperable malignant disease and bouginage in cases of stenosis Esophagoscopy is often a lifesaving procedure in cases of foreign body obstruction Generalized disorders such as pellagra sclero

derma, myasthenia gravis, poliomyelitis, bulbar palsy, and specific and nonspecific infectious diseases may involve the esophagus. Esophagitis is far more common than is generally supposed and may develop into inflammatory or fibrous stenosis. True peptic ulcer of the esophagus is not very common, and seems to be related to esophagitis, short esophagus, hiatus hernia or duodenal ulcer. Agranulocytic esophagitis, caused by drug sensitivity, is sometimes found. When in addition to these diseases one considers trauma, extrinsic pressure, congenital stenosis, emotional disorders of swallowing, and esophageal varices, one must realize the importance of the esophagus.

The ear, nose and throat specialist has traditionally regarded the esophagus as part of his realm. Nowadays, however, the thoracic surgeon, the gastroenterologist, the endoscopist, and often the laryngologist are interested in esophagoscopy. These specialists should therefore keep themselves informed about diseases of the esophagus and about the best methods of diagnosis and treatment. They should know, for example, when either bronchoscopy or esophagoscopy is advisable in carcinoma of the esophagus, how and when to carry out bouginage for benign stenosis and achalasia, and whether to refer such cases to the surgeon.

The endoscopist, be he gastroenterologist, laryngologist, thoracic surgeon, or solely endoscopist, must be much more than a mere passer of tubes. He must see the patient as a whole and be able to interpret findings on the basis of all available information about the patient. He must, through association with internists and surgeons, keep informed about methods of diagnosis and treatment. To quote from Churchill's foreword to Benedict's *Endoscopy* (1): "Someone has said that endoscopy is merely the art of looking through a tube. Could anyone be so rash as to say this of the great science of astronomy? Of course the technics of both astronomy and endoscopy can be brought to the carnival level where one may drop a quarter in the slot and look at the moon, or for the same price watch the sword swallower pass a strip of metal down his esophagus. . . ." Churchill concludes that endoscopy has been

molded into a productive and significant area of medicine by making the subject matter of the area first and foremost the patient and not merely the technical skill of taking a look at his insides

When a patient with an established diagnosis of specific esophageal disease presents himself for treatment the doctor must decide whether the problem requires medical endoscopic or surgical treatment An uncomplicated peptic ulcer of the esophagus with no stenosis and no hemorrhage may be successfully treated by medical procedures however if stenosis occurs the endoscopist should be called upon for bouginage and if the stenosis does not respond to bouginage surgery must be considered Esophageal ulcer may cause massive hemorrhage in which case surgery must be considered after the first episode and becomes mandatory after the second Carcinoma of the esophagus is a medical problem when there is no obstruction and operation is contraindicated because of age widespread metastases or other serious complications If the esophagus becomes obstructed owing to the disease to edema or to fibrosis following x ray treatment the endoscopist should be called upon for bouginage if this is not successful the general surgeon may be consulted Most problems of esophageal surgery however require a thoracic approach hence they are best handled by a thoracic surgeon or by a general surgeon who has had a great deal of experience in thoracic surgery

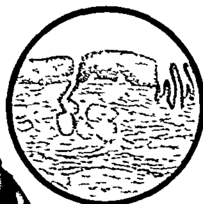
In a large hospital there is much to be gained by discussing esophageal problems before a group consisting of gastroenterologist thoracic surgeon radiologist and endoscopist Often many internists and surgeons are present at such meetings together with house staff visitors and graduate and undergraduate students Patients are presented and their cases described in some detail by the house staff x rays are shown by the radiologist endoscopic findings are explained by the endoscopist and advice is given by the group concerning further study and treatment

It should be remembered that esophageal surgery though firmly established is still in a developmental phase and that only rarely

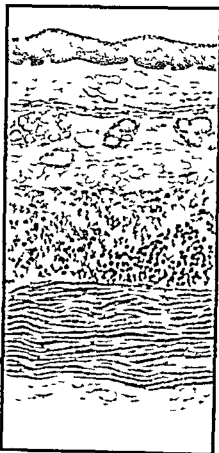
will the results of any single series be statistically valid We have attempted to review our experience in the light of reports from colleagues and to present surgical technics which we believe are currently the safest and most effective

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ESOPHAGOGASTRIC JUNCTION



ESOPHAGUS THROUGH MIDREGION

Fig. 1 The esophagus – structures, relations, histology. (Courtesy Fletcher D. Woodward, M.D., Clinical Symposia 13, 195[illegible].)



DISPLACEMENT OF ESOPHAGUS BY ENLARGED HEART

DISPLACEMENT OF ESOPHAGUS BY ANEURYSM OF AORTA





F. Netter M.D.
CCIBA

PLATE II. Displacement of the esophagus. (Courtesy Fletcher D. Woodward, M.D., Clinical Symposia 8:3, 1956.)

Anatomy of the Esophagus

2

This chapter reviews only those fundamentals of anatomy and embryology essential for a useful understanding of esophageal development and structure. The detailed anatomical knowledge required for carrying out specific surgical procedures or for understanding particular esophageal disturbances will be treated at greater length in subsequent chapters.

Embryology

The esophagus develops from the primitive foregut together with the pharynx and the stomach.

In a three week embryo the esophagus is represented by a mere constriction between pharynx and stomach. When the neck differentiates at the beginning of the second month the esophagus undergoes rapid elongation owing to the development of the lungs and pleural cavities which process pushes the stomach backwards into the abdominal cavity.

The diaphragmatic septum transversum originates from the cer

vical mesoderm and descends caudally with the phrenic nerve. During its development the stomach descends to overtake and pass the septum transversum, entering the abdomen before the lateral diaphragmatic components fuse with the septum transversum. If gastric descent is slow the stomach may become trapped in the thorax, and a congenitally short esophagus with thoracic stomach results. This problem and its ramifications will be discussed in detail in Chapter 9.

The esophagus and trachea become differentiated during the third and fourth weeks as a result of the ingrowth of two lateral septa which fuse and thus form the posterior or membranous wall of the trachea and the anterior wall of the esophagus. The most caudad portions of these septa fuse last, at the level of tracheal bifurcation. The development of esophagotracheal fistula (Chapter 26) owing to abnormal fusion is therefore readily understandable.

After trachea and esophagus have been separated by the fusion of the lateral septa, the esophageal lumen is almost obliterated by proliferation of the epithelial lining. Some workers believe that complete obliteration occurs. However, others maintain that the original lumen never disappears. These irregularities in the esophageal epithelium have been thought to cause atresia, stenosis, and possibly diverticula, but direct evidence is lacking. Large vacuoles then appear in the thickened epithelium, and hence in cross section the esophagus may appear to have several lumina. Subsequently the lumen enlarges owing to coalescence of these vacuoles. Failure of some vacuoles to coalesce may explain some esophageal cysts (Chapter 18). The mucosa of the esophagus, which is at first of simple columnar form, goes through a transient stage of ciliation before it attains its final stratified squamous condition. At this early stage the epithelium of the esophagus is thrown into longitudinal folds and on cross section the lumen is cruciform.

General Features

The adult esophagus (Plate I) is a soft-walled movable muscular tube approximately 25 cm (10 inches) in length. In the infant it is about one half this length. When empty it lies flattened anteroposteriorly, and when distended it is irregularly cylindrical in shape. It begins in the neck at the lower border of the cricoid cartilage opposite the sixth cervical vertebra. Its upper boundary is marked on the outside by the lower border of the inferior constrictor muscle. As it descends through the mediastinum the esophagus describes a gentle S-curve in the anteroposterior plane. In its first or cervical portion it tends to lie a little to the left. Accordingly, the surgical approach to lesions in the neck (*e.g.*, diverticulum) is made through the left side of the neck. In its second or upper thoracic portion the gullet swings gently to the right, more to the right of than anterior to the thoracic aorta. For this reason many high lesions are most easily reached through the right upper thorax. At this level the esophagus is immediately adjacent to the right parietal pleura behind the pulmonary hilus and hence is vulnerable to injury during pulmonary surgery. In its lowermost portion it again swings to the left behind the pericardial sac and then anterior to the aorta. It passes through the diaphragmatic hiatus at the level of the tenth thoracic vertebra. Accordingly lesions in this area are best approached through a left lower thoracotomy. It also shows a general anteroposterior curve from above downwards in conformance with the curve of the vertebral column upon which it lies.

The thoracic duct lies between the esophagus and the prevertebral fascia in the right lower posterior mediastinum. It crosses over to the left at the level of the fourth thoracic vertebra and continues its course up into the neck.

The longitudinal and circular musculature of the upper third of the esophagus is striated; that of the lower two-thirds is nonstriated. This may be of significance in the etiology of achalasia (Chapter 14).



FIGURE 1 X ray appearance in esophageal obstruction due to hypertrophy of the cricopharyngeus Note barium retention in valleculae and pyriform sinuses

The organ narrows at three points (1) at its origin, owing to the action of the cricopharyngeus (Figs 1, 2), (2) at a point approximately 7 cm below the cricopharyngeus, where the esophagus passes behind and to the right of the aortic arch, continuing into the posterior mediastinum, where at a point 4 cm below the arch it passes behind the left main bronchus (the resultant aortic and bronchial narrowing may be observed at the time of esophagoscopy and are of some importance as landmarks), and (3) at the point at which the esophagus passes through the diaphragm It usually continues into the abdomen for approximately 1 cm before joining the stomach at the cardia



FIGURE 2 X-ray appearance showing filling defect due to hypertrophy of cricopharyngeus

A chest x-ray made in the right anterior oblique position demonstrates the so called esophageal triangle which the distal half of the esophagus fills (Fig 3, Plate II) This triangle is bounded by the heart anteriorly, by the descending aorta posteriorly, and by the diaphragm inferiorly

STRUCTURE

The esophageal wall consists of four coats (1) adventitia (2) muscularis (3) submucosa and (4) mucosa. The esophagus differs



FIGURE 3 X-ray appearance of normal esophagus passing through the esophageal triangle The normal folds are well shown by relief technic Constrictions due to the cricopharyngeus aorta and left main bronchus can be seen It is also obvious in this view how dilation of the left auricle could cause an extrinsic filling defect of the esophagus. This is usually best seen in a left lateral projection during a barium swallow

from the rest of the gastrointestinal tract in that it has no serosal covering An outer fibrous sheath takes the place of the serosa This adventitial layer contains the vagi and many of the larger blood vessels

The muscular coat is composed of outer longitudinal and inner circular fibers Approximately 4 cm below the cricoid the posterior longitudinal fibers diverge into two bundles which pass upward and forward and insert on the cricoid cartilage A V-shaped defect is left between the diverging bundles this is filled by the circular muscle fibers and is reinforced by some decussating longitudinal fibers and, superiorly by overlapping cricopharyngeus fibers This V shaped defect is erroneously considered to be the site or origin of diverticula Diverticula (see Chapter 16) actually originate from the triangular space (Plate III) Between the circular and longitudinal muscles is the intermuscular septum containing Auerbach s plexus and a vascular network The circular musculature is relatively weak and although a longitudinal closure can be satisfactorily effected a transverse closure as in end to-end anastomosis may be precarious Accordingly sutures should be placed as horizontal mattresses in this layer and chief reliance should be placed on the mucosal layer

The submucosa contains many mucus glands and blood vessels Meissner s plexus and a rich system of lymphatics

The mucosa is pinkish at the upper end of the normal esophagus and gradually becomes a pearly white until there is an abrupt change at the cardia where the red irregular gastric mucosa begins The esophageal mucosa consists of (1) a thick layer of stratified squamous epithelium ( ) a thick layer of connective tissue and (3) a strong muscularis mucosa This last is the critical layer for the placement of sutures in esophageal surgery

Arterial Supply

In the neck the esophagus is supplied (Plate IV) by the superior esophageal arteries which arise from the inferior thyroid artery

and the highest intercostal arteries In the thorax the middle esophageal arteries originate directly from the thoracic aorta and from some bronchial and intercostal arteries In its lowest portion the esophagus is supplied by the inferior esophageal arteries, which arise from the abdominal aorta, the inferior phrenic artery, and the left gastric artery

The portion of the esophagus adjacent to the tracheal bifurcation usually has the richest blood supply The lower segment of the thoracic esophagus is the next richest The upper half of the thoracic esophagus just below the region of the tracheal bifurcation is believed to have the poorest blood supply, the surgeon should keep this in mind when planning an esophageal anastomosis at this level below the aortic arch

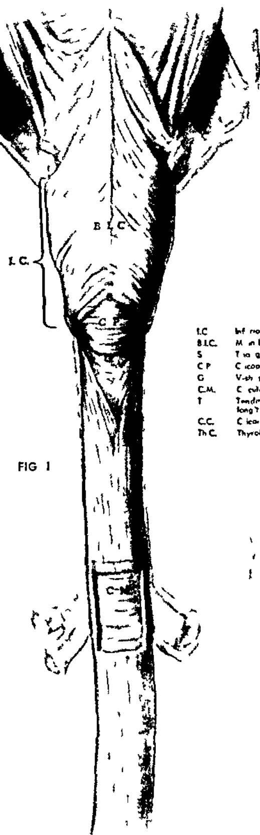
Venous Channels

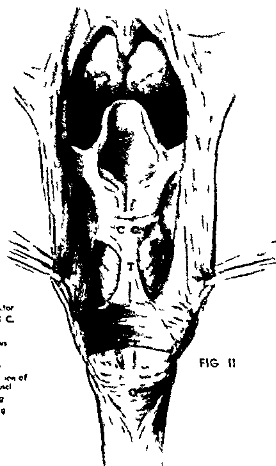
The venous distribution (Plate IV) corresponds to the arterial supply The superior esophageal veins drain into the subclavian and thyroid veins The middle esophageal veins drain into the azygos and hemiazygos veins The drainage of the lower thoracic and abdominal esophagus takes place through the coronary, left gastric and splenic veins which are tributaries of the portal vein In this fashion a communication is established between the portal and systemic circulation to which we will refer again in the chapter on esophageal varices and portal hypertension (Chapter 17)

Lymphatics

There are two lymphatic plexuses (Plate V) in the esophagus one in the submucosal layer which is continuous with that of the pharynx and another which lies between the muscle layers

It has been generally accepted that there is a regional lymphatic drainage system for the esophagus However, the collecting channels in the submucosa run for long distances before perforating





I.C	Inf rior Constrictor
B.I.C.	M in body of I. C.
S	T ia gular sp
C P	C icopho yngeus
G	V-sh ped g p
C.M.	C cular muscle
T	Tendinous ins ion of long't dinal muscl
C.C.	C icoid rtil g
Th C.	Thyroid c rtil g

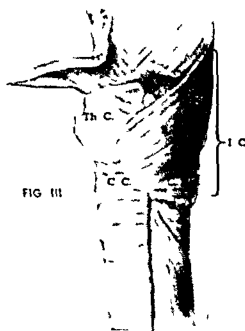


Plate III The musculature of the esophagus (Courtesy Fletcher D Woodward M.D., Clinical Symposia 8 3 1956.)

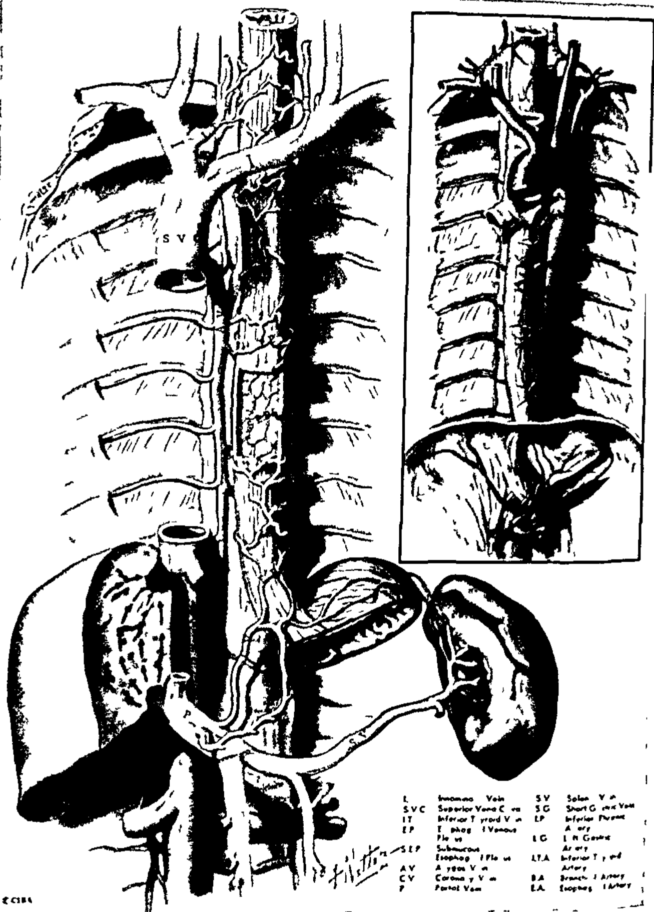


Plate IV Blood supply of the esophagus. (Courtesy Fletcher D. Woodward, M.D., Clinical Symposia 8:3, 1956.)

the wall and draining into a group of nodes Accordingly the concept of a segmental node distribution is open to question

Nerves

The esophagus has a dual nerve supply (Plate V) Parasympathetic fibers are supplied by the vagus nerve The recurrent laryngeal nerves supply the upper portion and the vagal trunks supply the remainder of the esophagus directly sending parasympathetic *preganglionic fibers to the musculature* Sympathetic fibers are supplied in the neck by the superior and inferior cervical ganglia in the thorax by the upper thoracic and splanchnic nerves and in the abdomen by the celiac ganglion These probably consist largely of postganglionic motor fibers As previously mentioned these nerves form plexi within the esophageal wall The ganglia associated with these plexi are vagal they represent the origin of postganglionic sympathetic fibers The afferent pathways are not known

Surrounding Structures

The esophageal hiatus in the diaphragm is formed by two anastomotic muscular bundles which originate from the ventral portion of the crural pillars decussate surround the esophagus and enter into the tendinous diaphragm Ventral to the esophagus these fibers are reinforced by others of the diaphragm The hiatal musculature is the most important means of fixation of the esophagus The latter loops under the hiatal muscle sling which pulls backward and to the right, maintaining and accentuating the physiologic angulation between esophagus and stomach The esophagus and vagus nerves pass through the hiatus surrounded by loose reticular tissue which permits sliding and contains the paracardial lymph nodes Above and external to this tissue a layer of connective tissue rich in elastic fibers is attached to the fascia propria of the esophagus

and below is fixed to the margins of the hiatus and is continuous with the subdiaphragmatic and transversalis fascia This phrenico-esophageal ligament constitutes another means of elastic fixation A third less important means is the peritoneal reflection

Physiology of the Esophagus

3

Physiologic investigations of the esophagus are essentially studies of swallowing and of the mechanics of transport of nutrition from the pharynx to the stomach

The initial stages of swallowing are oral and voluntary they consist in forming a bolus and delivering it into the posterior pharynx The resultant simulation of the posterior wall of the pharynx and of the soft palate tongue and epiglottis initiates a series of involuntary coordinated reflexes which propel the bolus into the upper esophagus and initiate the esophageal phase of deglutition

The cricopharyngeus which guards the entrance to the esophagus is normally in a state of moderate tonic contraction this contraction except when vomiting takes place prevents regurgitation of food and the entry of esophageal secretions into the mouth This tonic contraction is quite evident to the esophagoscopist The cricopharyngeal sphincter is a powerful one it is always closed except when deglutition or ejection is actually occurring The normal tone of the cricopharyngeus is quite variable in different persons

and this variability is the chief factor in the ease or difficulty of carrying out esophagoscopy.

Liquids and semi-solids pass down the esophagus whether or not peristalsis takes place (4). The peristalsis initiated by swallowing, however, continues in its usual fashion.

Normal peristalsis in the esophagus has been shown to consist of two types of waves:

(1) a primary wave initiated by deglutition starting at the upper end of the esophagus proceeding without interruption to the cardia, and providing the chief propulsive force for moving a bolus, and (2) a secondary wave which is initiated not primarily by deglutition but secondarily by distention of the esophagus. These waves are absent in the upper third of the esophagus (5), begin at about the level of the aortic arch, and are less powerful than primary waves (6). Liquids and semi-solids pass down the esophagus whether or not peristalsis takes place. The peristalsis initiated by swallowing, however, continues in its usual fashion.

Tertiary waves, reverse peristalsis, and other contractions have been described, but these are probably abnormal and will be considered in detail with the esophageal diseases wherein they are present.

The transit of a bolus of food to the end of the esophagus requires about 5 seconds. At this level there is usually a momentary delay until the cardiac sphincter relaxes and permits the passage of the food into the stomach. The normal cardia then quickly closes and prevents regurgitation of gastric contents back into the esophagus.

The normal propagation of peristaltic waves and their coordination with cardiac relaxation are probably dependent on proper synchronization of the action of the sympathetic and parasympathetic nerves. The vagus or parasympathetic nerves are probably responsible for the maintenance of the normal tone of the esophagus. Peristalsis in the esophagus is mediated through its extrinsic motor nerves (the vagi), and it does not occur when these are divided or when their centers in the brain are paralyzed, as during anesthesia

Animal experiments suggest that the functions of the parasympathetic nerves are motor in the esophagus and inhibitory at the cardia and that the sympathetic nerves act antagonistically Such clear-cut antagonistic action of vagi and sympathetics has not been convincingly demonstrated in man This problem will be considered further in Chapter 14

The cardiac sphincter of the lower esophagus remains as difficult to find as the cricopharyngeus is accessible The term cardia has no precise anatomic connotation It was coined by Galen who believed that diseases of the lower esophagus simulated disorders of the heart It is doubtful that an anatomic cardiac sphincter exists even though the circular muscle coat is slightly thicker at the lower end X ray studies have shown that there is a physiologic sphincter located 1 to 3 cm above the level of the diaphragm This sphincter can usually be seen by esophagoscopy in fact it may be so tightly closed that it prevents the passage of the esophagoscope into the stomach It relaxes under general anesthesia This constricting area has been termed by some the lower esophageal sphincter It may occasionally be confused with a lower esophageal ring (see Chapter 13)

The closure of the cardia is probably the resultant of three main anatomic and physiologic factors

(1) The increased negative pressure in the chest and the increased positive pressure in the abdomen combine to cause the ascent of the cardia through the hiatus The right crus of the diaphragm kinks and compresses the gullet and thus tends to prevent regurgitation (This may be seen on inspection)

(2) The acid gastric contents may stimulate the physiologic cardiac sphincter to contract

(3) The cardiac valves The oblique insertion of the esophagus on the stomach forms the so called valve of Guberoff The angle is formed by the left wall of the esophagus and the right wall of the gastric fundus This angle is maintained partly by the contraction of a string of muscle fibers known as the muscle loop of Willis partly by the ballooning of the gastric fundus by the normal air

bubble and partly by the gastrophrenic ligament Another valve effect results from the leftward turning and partial twisting of the terminal portion of the esophagus immediately below the diaphragm

The diaphragmatico-esophageal ligament may also play a role in competence and deglutition Laimer (3) originally described this structure as a continuation of the transversalis fascia on the undersurface of the diaphragm It divides at the hiatus into (1) an ascending part that extends upwards and attaches to the lower esophageal musculature and (2) a descending limb attaching at the cardia *This structure is antagonistic to the longitudinal musculature* and prevents the esophagus from pulling the stomach into the chest during deglutition

Intrinsic movements Normally the esophagus is a lax muscular tube (Plate VI #1, #2, see also Figure 3) Peristalsis occurs only after a distinct act of swallowing Peristaltic waves in the esophagus are best observed at x-ray examination, they cannot be seen at esophagoscopy *The reverse peristalsis which occurs in the* esophagus during belching regurgitation, and vomiting also may be observed by x-ray but not by esophagoscopy *Another intrinsic* movement of the esophagus is spasm This may occur throughout the length of the esophagus and likewise can be well demonstrated by x-ray examination but not by esophagoscopy Such spasm is known as esophagospasm this is discussed in detail in Chapter 15

Extrinsic movements Normal respiration causes little observable change in the lumen of the esophagus However, during deep *inspiration the esophagoscopist notes a widening of the lumen* which may involve the cardiac orifice Such a widened lumen may greatly help the examiner by permitting *a better view of a stric*ture a carcinoma or a foreign body Better forceps space for removal of a biopsy specimen or a foreign body may be obtained by asking the patient to take a deep breath Such a deep inspiration also may help the examiner to find the direction of the lumen and to pass the esophagoscope deeper into the esophagus or through the cardiac orifice into the stomach

The transmitted pulsations of the heart and aorta against the esophageal wall are readily observed during esophagoscopy They are significant only as landmarks

Aerophagia (air swallowing) is a well recognized entity which occurs in nervous persons The x-ray appearance is shown in Figure 4 The woman whose esophagus is shown in the illustrations swallowed air and stored it in the esophagus by keeping the cardiac and pharyngeal constrictors closed The esophagus dilates to at least 3 times its normal diameter measuring about 4½ cm Only a small amount of air reaches the stomach After the esoph-

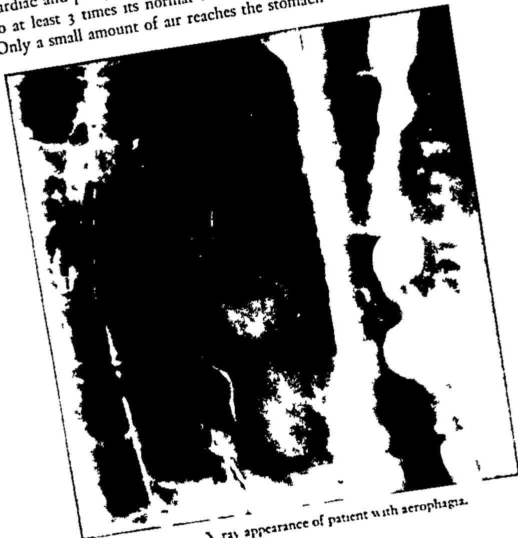


Figure 4 X ray appearance of patient with aerophagia.

agus is filled with air the patient is able to swallow the barium and let it pass into the stomach without emptying the gas from the esophagus After a time the esophagus is emptied with a sudden belch and the process begins again

Cricopharyngeal hypertrophy In rare cases the powerful cricopharyngeal sphincter becomes hypertrophied and this causes some esophageal obstruction (1) Such a case is shown in Figures 1 and 2 The patient whose x-rays are illustrated has been partially relieved by esophagoscopy and bouginage Rogers (5) reported a patient with dysphagia associated with cricopharyngeal spasm who was treated successfully by bilateral superior cervical ganglionectomy

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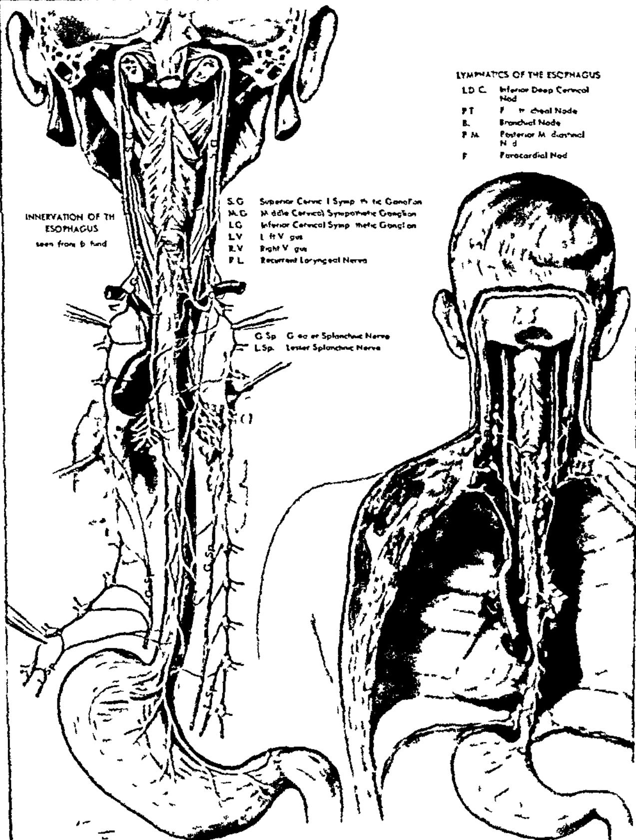


Plate V. Nerves and lymphatics of the esophagus. (Courtesy Fletcher D. Woodward, M.D. Clinical Symposia [illegible])








PLATE VI Esophagoscopy

1 Normal esophagus at about the mid thoracic portion at the end of expiration Note smooth pale pink mucosa Esophageal lumen contracts down and often disappears completely at the end of expiration

2 Normal esophagus slightly lower down during inspiration The normal esophageal lumen expands markedly during inspiration

3 Multiple telangiectases of the esophagus in a child two years old whose chief symptom had been hematemesis

4 Marked esophagitis lower end of the esophagus in a middle aged man *whose chief complaints were substernal burning and some difficulty in* swallowing solid food Esophagoscopic biopsy confirmed chronic esophagitis Bland diet and bouginage resulted in marked improvement

5 Inflamed edematous easily bleeding gastric rugae in a hiatus hernia Esophagoscopic biopsy showed chronic gastritis in the herniated portion of the stomach Esophagoscopy and bouginage gave considerable relief

6 Appearance of benign stricture of the esophagus as seen at the time of esophagoscopy The lumen of the esophagus is contracted to a diameter of approximately only 2 mm There is smooth concentric narrowing with slight inflammation This patient had had a subtotal resection for duodenal ulcer followed by regurgitation and esophagitis Bougies sizes 8 to 24 were passed through the esophagoscope and the patient was relieved

FUNDAMENTALS OF DIAGNOSIS

4

CLINICAL MANIFESTATIONS

A thorough clinical history is essential as this may reveal the first indication of esophageal disease In fact the diagnosis may be missed if the physician does not inquire in detail about dysphagia odynophagia heartburn substernal pain gas nausea vomiting regurgitation hematemesis melena anorexia loss of weight excessive salivation cough noisy deglutition lump in throat hoarseness and choking and gagging It should also be borne in mind that fever pulmonary emphysema and shock may be encountered in perforated or ruptured esophagus

Dysphagia means difficulty in swallowing We must distinguish between dysphagia as difficulty in the act of swallowing and dysphagia as a feeling that food is stuck in the esophagus Some patients deny any difficulty in the act of swallowing but admit that food seems to get stuck before it reaches the stomach This is a correct and important observation Difficulty in the act of swallowing is less frequent than the feeling that food is stuck in the esophagus The former is likely to indicate myasthenia gravis or

some type of paralytic dysphagia such as occurs in poliomyelitis bulbar palsy, medullary infarction, diphtheritic neuritis infectious polyneuritis with paralysis of the tenth nerve, and nerve trauma with resultant glossopharyngeal paralysis

The feeling that food is stuck in the esophagus is often one of the earliest and most important symptoms of esophageal disease It *is usually a concomitant of carcinoma, benign inflammatory steno*sis, congenital atresia, caustic stenosis, achalasia and obstruction due to web or foreign body It may be present in esophagitis esophageal ulcer, hiatus hernia, diverticulum, benign tumor and specific infectious diseases This type of dysphagia also may be caused by extrinsic pressure on the esophagus, and it may be present in pellagra and scleroderma

Emotional dysphagia, sometimes termed globus hystericus hysterical dysphagia, or functional or psychosomatic dysphagia, is rare

Odynphagia means painful deglutition It is sometimes found in esophageal ulcer, severe esophagitis benign stenosis, and foreign body obstruction

Heartburn is too common a symptom to be in any way specific for esophageal disease It may occur in esophagitis, benign stenosis esophageal ulcer, hiatus hernia caustic stenosis carcinoma, and achalasia

Substernal pain also is not specific for esophageal disease, but should lead one to consider esophagitis benign stenosis esophageal ulcer hiatus hernia, foreign body obstruction tumor and achalasia The pain of esophageal disease may be very much like that of angina pectoris Obviously the differentiation is of very great importance

Gas Patients with esophagitis, benign stenosis esophageal ulcer, hiatus hernia or achalasia may complain of gas but it is a *non* specific complaint Inability to belch is sometimes a rather distress*ing accompaniment* of lower esophageal obstruction patients suffering from it are often greatly pleased to be able to belch normally again after bouginage

Nausea is a symptom which may be present in almost any disease of the esophagus but it occurs in so many other disorders that it is not significantly related to the esophagus

Vomiting is a common symptom of many diseases it leads one to think first of the stomach for by definition to vomit is to eject from the stomach through the mouth' Real vomiting therefore cannot be primarily due to esophageal disease Vomiting implies a muscular contraction of the stomach wall with reverse peristalsis Reverse peristalsis of the esophagus may occur but it leads to regurgitation rather than vomiting

Regurgitation is one of the commonest symptoms of esophageal obstruction It is an overflowing of secretions fluid and food material from the esophagus through the mouth Such material may be expectorated or may be aspirated into the trachea with resultant tracheobronchial and pulmonary infection Overflowing of such material at night often results in drooling It is a prominent symptom of achalasia, especially when the condition is of long standing It also occurs in other types of benign and malignant obstruction

Hematemesis Strictly speaking this means the vomiting of blood In patients with esophageal varices massive regurgitation of blood is common since a large amount of blood may accumulate in the stomach from these varices Bleeding also may be caused by esophagitis esophageal ulcer hiatus hernia benign or malignant tumor and occasionally by a foreign body or diverticulum Such bleeding usually occurs in relatively small amounts in which case blood may be found in the stool or in regurgitated esophageal material (See Plate VI #3)

Melena may occur owing to esophageal varices or esophageal ulcer Other diseases of the esophagus such as esophagitis hiatus hernia or tumor may produce occult blood in the stool but they seldom produce melena Moreover since many diseases of the stomach and intestine may give rise to melena these possible sources of bleeding also should be investigated

Anorexia is a fairly common complaint in esophageal carcinoma

It occasionally occurs in esophageal obstruction due to other causes Since it is also found in carcinoma of the stomach and in many other conditions it does not conclusively indicate esophageal disease

Loss of weight is another nonspecific symptom of many disorders It is not significant in the early diagnosis of carcinoma anywhere in the body since it usually occurs late in the disease no matter where the primary tumor is Loss of weight due to limited food intake occurs in all types of esophageal obstruction it is not diagnostic of carcinoma of the esophagus

Excessive salivation is commonly seen in esophageal carcinoma but also occurs in other types of esophageal obstruction

Cough when it occurs in esophageal disease is usually due to aspiration into the larynx or the tracheobronchial tree of material regurgitated from the esophagus Such aspiration may occur in achalasia and it frequently takes place at night when the patient is recumbent and asleep It occurs much less commonly in other types of esophageal obstruction Fistulous communication between esophagus and tracheobronchial tree also is a cause of cough

Noisy deglutition is commonly found in hypopharyngeal or esophageal diverticulum but may occur in elderly people in the absence of demonstrable disease

Lump in throat When a patient makes this complaint one must look for everything but may find nothing Such a complaint must be investigated by x ray laryngoscopy pharyngoscopy, bronchoscopy and esophagoscopy The symptom may be due to tumor but it often seems to be of functional origin and is not infrequently due to cancerphobia

Hoarseness often indicates intrinsic laryngeal disease but also occurs in paralysis of the recurrent laryngeal nerve due to pressure Carcinoma of the esophagus with invasion of the recurrent laryngeal nerve is a fairly common cause of hoarseness

Choking and gagging occur primarily as a result of lesions of the larynx and pharynx but sometimes secondarily owing to esophageal obstruction with aspiration of esophageal contents

Radiology

In any case in which there is the slightest suspicion of esophageal disease x ray examination of the esophagus by an expert radiologist is essential Unfortunately there are some radiologists who do not examine the esophagus with sufficient care and thus lesions may be missed

This book is not intended to teach x ray technic but it should be pointed out that a good x ray study of the esophagus ought to include investigation of the pharynx and pyriform sinuses the motility of the vocal cords the action of the cricopharyngeus the appearance of the esophageal mucosa the nature of the peristaltic wave the speed of barium passage the appearance of the cardiac orifice and any evidence of hiatus hernia The Valsalva maneuver may be helpful Spot films are often essential and during fluoroscopy the patient must be rotated in various positions —anteroposterior oblique and lateral both standing and recumbent The double-contrast method (contrasting the barium visualized mucosa with swallowed air) is useful The flexibility of the esophagus should be investigated any rigidity indicates disease Intrinsic defects and abnormal extrinsic pressure must be carefully studied

Although a negative x-ray examination goes a long way toward assuring the clinician and the patient that there is no disease in the esophagus the diagnostic value of the method is limited notably in cases of esophagitis web varices and non opaque foreign bodies Moreover it cannot always be relied upon to differentiate benign and malignant esophageal obstruction Therefore although x ray examination should usually be the first procedure esophagoscopy is also of very great importance in the diagnosis and treatment of esophageal disease

Esophagoscopy

Esophagoscopy (Chapter 5) is generally indicated whenever esophageal disease is suspected This is particularly true in the following cases

(1) If the radiologist is in doubt For example in examining a smooth stenosing lesion of the esophagus the radiologist may be unable to differentiate spasm, benign stenosis, and carcinoma Esophagoscopy with biopsy should result in a positive diagnosis

(2) If a histologic diagnosis is desirable Even when the roentgenogram shows a fairly typical picture of carcinoma it is important to establish diagnosis through biopsy before deciding on thoracotomy or x-ray therapy

(3) If x-rays are negative but esophageal symptoms persist Esophagoscopy may reveal esophageal web, esophagitis, or esophageal erosions

The more expert the radiologist is in the diagnosis of esophageal disease the less likely is it that the endoscopist will change or add to the x-ray diagnosis However, foreign body in the esophagus often requires esophagoscopy for diagnosis as well as for removal Esophagoscopy is often indicated in the diagnosis and treatment of benign peptic stenosis caustic stenosis web and achalasia Esophageal varices are sometimes seen by esophagoscopy when they cannot be visualized by radiology

The basic technics of esophagoscopy are discussed in Chapter 5 In performing esophagoscopy as a diagnostic procedure it should be remembered that while it is usually possible to see all parts of the esophagus with an open tube esophagoscope foreign bodies may occasionally be lost in the folds particularly in the region of the cricoid cartilage In megaesophagus as seen in long standing achalasia the lumen of the esophagus is so large and may be so much obscured by food material that complete investigation is rather difficult in such cases examination should be preceded by esophageal lavage (see Preparation of Patient page 40) An ade-

quate specimen for biopsy should be obtained in every case in which careful endoscopic inspection indicates the possibility of carcinoma. One negative biopsy is not sufficient to exclude carcinoma. For the differential diagnosis of benign inflammatory stenosis and smooth annular carcinoma the tissue must be obtained from deep within the lumen of the stenosed area but not so deep as to be beyond a small carcinoma.

Many benign stenoses are amenable to bougienage which is usually begun at the time of the first esophagoscopy. Similarly in achalasia if esophagoscopy is indicated for differential diagnosis bougienage can be performed at the time of the first esophagoscopy and further dilatations can be carried out later in the office or out patient department.

Bronchoscopy

Bronchoscopy should be performed in all cases of carcinoma of the esophagus near or above the carina in order to determine whether the tumor has invaded the tracheobronchial tree and whether there is paralysis of one or both vocal cords. If the tumor has actually invaded the trachea or bronchus it is surely inoperable. The finding of metastatic carcinomatous glands with paralysis of the recurrent laryngeal nerve is evidence of probable inoperability.

Cytology

Cytologic study of secretions aspirated from the esophagus at the time of esophagoscopy or independently obtained by Levine tube suction has proved to be an important addition to our diagnostic methods. Ordinarily an adequate biopsy specimen for carcinoma diagnosis can be obtained at the time of esophagoscopy but occasionally — especially when there is megaesophagus or when the malignant obstruction is unusually smooth — a small carcinoma may be missed completely because the biopsy forceps are directed to a non malignant area.

T A I a a -7-year-old white man was admitted in 193- with the complaint of difficulty in swallowing of - years duration He had been recently worrying considerably about financial matters and poor working conditions the dysphagia was worse at such times

X ray examination showed almost complete obstruction to the passage of barium at the cardiac orifice and the esophagus was considerably dilated above this point A diagnosis of achalasia was made The patient also had rheumatic heart disease with mitral stenosis

Mercury bouginage was carried out intermittently at the hospital and by the patient himself at home for 4 years In 1956 at the last bougínage at home he vomited bright red blood and reported immediately to the hospital X-ray examination showed findings consistent with longstanding obstruction of the distal portion of the esophagus as seen in achalasia The distal end of the dilated esophagus was somewhat irregular but there was no evidence of fistula formation Fluid and food in the esophagus prevented further investigation of the cause of obstruction

Esophagoscopy performed the next day revealed that the esophagus was dilated and contained a great deal of fluid The fluid was aspirated and a good view was obtained of the esophagus the mucosa was slightly reddened all the way to the cardiac orifice which also appeared normal There was no evidence of foreign body ulcer or carcinoma and no explanation of the bleeding was found Saline solution was introduced and secretions aspirated for cytologic study Esophageal washings obtained by Levine tube on two occasions were positive for cancer cells

In view of the positive cytologic findings it was planned to repeat esophagoscopy in order to confirm and localize the lesion if possible Unfortunately a second esophagoscopy was not carried out but instead the patient was operated upon immediately and a small (1 cm) carcinoma was found in the esophagus about 10 cm from the cardia The lesion was so small that it was difficult to locate even at operation Frozen sections of para esophageal lymph nodes indicated squamous-cell carcinoma Distal esophagectomy and proximal subtotal gastrectomy were performed with esophagogastric anastomosis The patient did not rally after the operation and died the next day Death was due to multiple pulmonary emboli

Comment This case is reported to show the value of cytologic examination in a patient with a dilated esophagus — a condition which always makes examination by x-ray and by esophagoscopy

particularly difficult not only because of the size of the organ but also because the esophagus is often filled with fluid

There are other examples of the value of cytologic examination in the diagnosis of carcinoma of the esophagus but they are less striking than the case described above For example, in one case carcinoma was believed to be the most likely diagnosis after x-ray examination but three esophagoscopies were performed before a positive biopsy specimen was obtained In this case it was the cytologic study which confirmed the diagnosis The carcinoma was small and the biopsy forceps presumably had been passed beyond the lesion and (as is very unusual) too deeply into the lumen beyond the stenosed area

A word of warning should be added regarding the possibility of false positive cytologic examinations Of course one should not proceed with major surgery on the basis of cell study alone A very definite x ray diagnosis of cancer plus positive findings from cytologic examination might be sufficient but the combination of a doubtful x ray examination and positive cytologic findings would be an indication for esophagoscopy and biopsy as is illustrated by the following case

W G a 54 year old white man entered the hospital for the first time in a semicomatose condition having been drinking heavily According to the statement of a brother the patient had been a chronic alcoholic for many years On the day before admission he had vomited blood and passed tarry stools

X ray examination showed no esophageal varices but revealed a slight deformity in the lower esophagus with what appeared to be a ring of calcification suggesting hemangioma The possibility of tumor could not be excluded A hiatus hernia was noted

Esophagoscopy performed a few days later showed the lower 5 cm of the esophagus to be considerably inflamed and eroded There were thick folds which at first suggested carcinoma but later appeared to be relatively smooth with numerous bleeding hemorrhagic erosions but no deep ulceration Since there had been no dysphagia bouginage was not necessary There was no evidence of hemangioma Several biopsies

mm -35 cm instrument being advisable for babies, the 6 mm, 35 cm for children up to three years, and the 7 mm, 45 cm for children from four to fourteen years of age

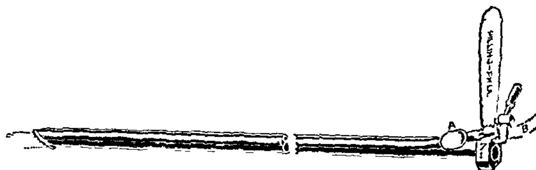


Figure 5 Standard full lumen 7 mm 45 cm Jackson esophagoscope equipped with Tucker thumb valve The 9 mm lumen is the one most frequently used for adults

The 45 cm esophagoscope reaches the cardiac orifice in most adults There is also an instrument 53 cm long which will pass through the cardiac orifice into the upper stomach, examination of these areas is often very important Larger esophagoscopes with lumens of 10 11, and 12 mm may be desirable when dilatation with larger bougies is necessary (see Bouginage, page 48) For example, the largest bougie that will pass through a 9 mm *esophagoscope is #26 French The 10 mm* 'scope will accommodate a #29 bougie, the 11 mm a #32, and the 12 mm a #35 The larger esophagoscopes are correspondingly *hard* to pass especially at the cricopharyngeus and should not be used except in well-relaxed, normal size adults If difficulty is encountered in passing the cricopharyngeus a #24 French bougie is lubricated and used as a guide through the esophagoscope to find the lumen after which the esophagoscope is passed into the esophagus being guided by the bougie

The Negus *esophagoscope* which *has proximal illumination* and a very large lumen is useful in passing the Souttar tube in certain cases of inoperable carcinoma of the esophagus *(Unfortunately* we have not had good results with the Souttar tube)

Suction

Most modern operating rooms are equipped with wall suction strong enough to divide into two parts one to connect with the aspirating channel in the esophagoscope and the other with an independent aspirator The latter is useful when it is necessary to reach specific areas of mucus or bloody secretion which lie a few centimeters beyond the tube mouth and also when the suction channel in the esophagoscope becomes plugged The independent aspirator should be 50 cm long for a 45 cm esophagoscope and 60 cm long for a 53 cm esophagoscope If wall suction is not available various types of suction pump may be used

Sponge carriers

These (Fig 6) are essential to hold tightly the small endoscopic sponges which must pass through the esophagoscope to enable

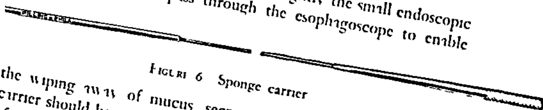


FIGURE 6 Sponge carrier

the wiping away of mucus secretions and blood The sponge carrier should be 50 cm or 60 cm long depending on the length of esophagoscope selected

Endoscopic sponges

These are made in four sizes to fit the lumen of the esophagoscope (4 mm 5 mm 7 mm 9 mm) They are best made from 3 inch closely woven gauze bandage Measuring plates are convenient but not essential The plate is placed crosswise on the bandage and a thread is pulled from the bandage with a pin in order to mark the exact width of unfolded sponge to be made The gauze rectangles are folded endwise and then once in the middle and strung together on a safety pin They are autoclaved before use

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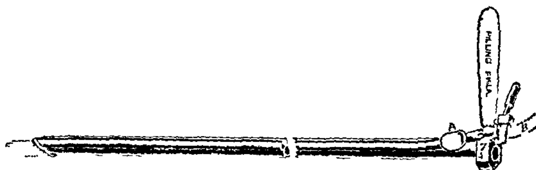


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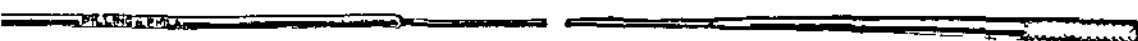


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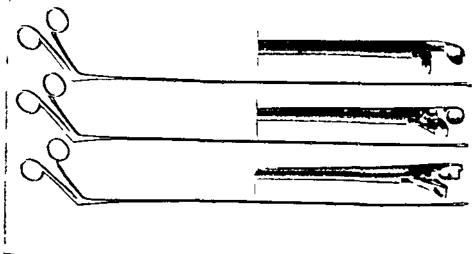


FIGURE 7 Various types of cutting forceps for tissue biopsy with close up views of the same forceps From top to bottom Jackson forward-cutting forceps Roberts s forward cutting forceps Roberts s side cutting forceps

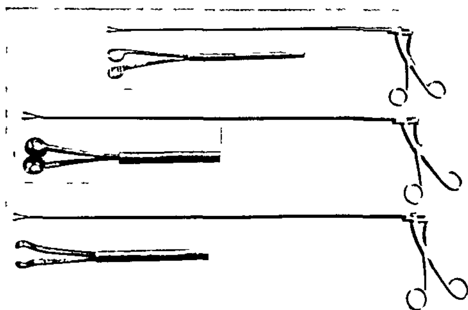


FIGURE 8 The three most useful forceps for removing foreign bodies with close up views of the same forceps These forceps come in different lengths as shown From top to bottom side curved forceps forceps for globular objects rotation forceps

Forceps

The most useful forceps for diagnostic esophagoscopy is the Jackson cutting forceps (Fig 7) for tissue biopsy A good bite with this forceps almost always gives an adequate specimen for microscopic diagnosis For foreign body removal the side curved forceps (Fig 8) is helpful the large-size forceps for globular objects enables one to extract pieces of meat without difficulty and the rotation forceps is valuable for grasping flat objects such as coins and flat pieces of bone

Esophageal bougies

Woven silk bougies (Fig 9) attached to a flexible steel shaft are essential for dilatation in stenosis of the esophagus They are graduated in size from #8 French to #35 French the smallest size being necessary for finding a tiny lumen the #26 being the



FIGURE 9 French esophageal bougie #17 with flexible silk woven tip fastened to a steel shaft

largest that will pass through the 9 mm esophagoscope They are important in dealing with benign inflammatory stenoses lye stric tures and webs and sometimes achalasia and carcinoma A large-size bougie is sometimes helpful in finding the lumen at the level of the cricopharyngeus

Operating table

Although a special operating table is not required it is most convenient to use one with a sliding top (Fig 10) which enables the patient to rest his head with his neck muscles relaxed until it is held by a trained assistant (see page 41) When the patient has to work himself up toward the head of the ordinary operating

Preparation of the Patient

The esophagus should be empty. Usually this condition can be effected by the patient's fasting for a few hours before the procedure. Small amounts of water may help to clean the esophagus. Even in cases of fairly complete obstruction due to carcinoma or benign stenosis, such preparation is usually adequate, and further cleansing can be secured by aspiration and sponging. In long-standing achalasia with megaesophagus, however, it may be advisable to cleanse the dilated esophagus by lavaging it, using a large old-fashioned stomach pump and a warm weak solution of sodium bicarbonate. Adequate sedation can be obtained by giving 0.1 gm or 0.2 gm of Nembutal two hours before the procedure and morphine sulfate (10 to 15 mg.) and atropine sulfate (0.4 mg.) subcutaneously about one-half hour beforehand. If the esophagus is badly obstructed, 0.12 gm. of sodium luminal given subcutaneously may be substituted for the Nembutal capsules.

The amount of sedation depends to some extent on the size and general make-up of the patient. A large, nervous, thick-set patient with a short neck, small mouth, and full set of teeth naturally requires more sedation than a small, phlegmatic person with a long flexible neck, large mouth, and removable dentures.

Anesthesia

Gargling with a 4 per cent solution of cocaine is usually adequate; this should be repeated once after a 5 minute interval. A 2 per cent solution of xylocaine is now routinely used in our clinic; it is considered less toxic than cocaine. Some patients, however, require general anesthesia. The risk of trauma to the delicate esophagus is so great in an uncooperative patient that general anesthesia is well worth while under such conditions. An experienced anesthesiologist can usually effect this with intravenously administered pentothal-anectine combined with intratracheal

intubation (to permit artificial respiration in case of bronchospasm)

Position of Patient

The patient lies flat on his back (see Operating Table, page 37) The head holder places a small towel around the patient s head grasping it firmly with his left hand near the vertex, and with his right hand slides the table top toward the operator During this maneuver the head end of the table automatically drops, and freedom for manipulation of the head is obtained The patient relaxes much better if he knows that his head is being carefully and firmly supported (See Figure 12)

The assistant who holds the patient s head must be comfortably seated on a stool about 18 inches high with his left foot elevated on an 8 inch box his left elbow supported by his left knee, and his left hand holding and controlling the patient s head The assistant then passes his right arm under the patient s neck, and with his

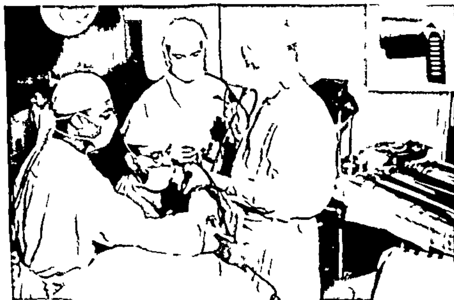


Figure 1 General view of operating room technic showing correct position for headholding bronchoscope in position to be introduced through the laryngoscope Headholding and position of patient is the same for introducing the esophagoscope but the laryngoscope is not used Inset shows bite block

right forefinger may insert a bite block (Fig 12) into the left corner of the patient's mouth For good exposure of the cricopharyngeus, the neck should be flexed on the body and the head extended on the neck, with chin up head thrust forward, *and shoulders flat on the table*

Postoperative Care

Routine orders after esophagoscopy are as follows (1) Aspergum stat at bedside (2) Cracked ice stat at bedside (3) Nothing else by mouth for at least 24 hours, and even then only liquids or foods ordered by the physician or house officer in charge *Aspergum is given freely for sore throat Cracked ice is used to moisten* the patient s mouth and provide him with a very small amount of fluid Nothing else is given by mouth until a responsible physician has checked the patient s temperature chart and his general condition Meanwhile the patient is kept comfortably hydrated immediately *after the esophagoscopy by the intravenous administration* of 1500 cc of 10 per cent dextrose in water If these precautions are observed, perforation should be discovered early, before further damage has been caused by the passage of liquids or solid food

Complications of Esophagoscopy

Perforation of the esophagus by an endoscopic instrument is the complication most dreaded by the endoscopist Since the esophagus is a thin musculomembranous tube, it is a delicate structure and may easily be perforated even by a skilled operator When the cricopharyngeus *fails to open readily, undue pressure on it by* the tip of the esophagoscope may cause a posterior perforation at that level by scraping the posterior wall of the esophagus against the body of the sixth cervical vertebra especially if exostoses are present (Fig 13) Such a complication is more likely to occur in *weak elderly women who have poor tissues and may be dehydrated* owing to esophageal obstruction *To avoid perforation* especially at that level one *should observe the following rules*



FIGURE 13 X ray appearance of marked cervical exostoses In this case it was not possible to pass the esophagoscope even under general anesthesia because of exostoses and stiff neck A rare situation but if the endoscopist had persisted perforation would have been almost certain.

(1) Extreme gentleness in all manipulations

(2) Thorough lubrication of the esophagoscope with mineral oil

(3) Adequate hydration of the patient, if necessary, by preoperative intravenous therapy

(4) Adequate preoperative sedation and cocainization

(5) Expert head holding

(6) If necessary, use a large bougie (#-4 French) as a guide in passing the esophagoscope

(7) Use of a smaller (8 mm or 7 mm) esophagoscope in weak or elderly patients

(8) Use of general anesthesia in nervous, hypertonic, or uncooperative patients

If these fundamentals are scrupulously observed, perforation should not occur Unfortunately, however, accidents occasionally happen and then the most important thing is to recognize them and treat them properly Perforation at the level of the cricopharyngeus may be obvious at the time of esophagoscopy, manifesting itself by slight bleeding and the posterior appearance of a dark cavity which leads nowhere The operator, however may be completely unaware that a perforation has occurred until several hours later when the temperature may rise to 100° F the throat may be very sore, it may be very painful to try to swallow saliva, foamy material may be spit up, and the neck may show subcutaneous emphysema and some stiffness X-ray examination shows air behind the esophagus If the usual routine postoperative orders (see page 42) have been observed, the patient should not be seriously ill and should respond to penicillin intravenous therapy and bed rest Nothing should be given by mouth After about one week the symptoms usually subside and the patient may safely begin drinking water in small amounts In rare instances a traumatic diverticulum forms at the site of perforation in which case surgical repair is indicated

Perforations of the middle and lower esophagus are less common but they are generally serious and may lead to mediastinitis and possible abscess formation If the diagnosis is not obvious the

roentgenologic demonstration of air in the mediastinum is usually conclusive

Minor perforations often respond to chemotherapy Major perforations require surgical intervention the type of surgery depending on the findings at exploratory thoracotomy (See Chapter 25)

Hemorrhage may occur during or after esophagoscopy The injection of sclerosing solution into varicose veins is likely to be attended with some bleeding Although this method of treatment is no longer used at the Massachusetts General Hospital it is still used in other parts of the United States The passage of bougies in patients with lye stricture severe esophagitis or esophageal ulcer usually causes slight bleeding and occasionally results in moderately severe hemorrhage Some minor bleeding always occurs after biopsy but this is seldom of any consequence

METHODS OF TREATMENT

6

DIETARY TREATMENT

When one thinks of the esophagus as a relatively narrow tube through which all food and drink must pass to reach the stomach, and when one considers the trauma to which it is subjected by unwise eating and drinking habits, it is obvious that one of the first essentials of good treatment is proper diet. Sometimes proper diet means that nothing should be given by mouth, as when the patient regurgitates or vomits everything and hence the esophagus *must* have complete rest for a few days, even before a diagnostic x-ray is undertaken. Under such conditions the patient must be maintained on intravenous therapy. At other times a proper diet means only bland liquids given in order that some nourishment may pass *an inflammatory stenosing lesion without trauma and without* sticking. (Some fruit juices, for example, may be irritating.) Obstructing tumors may require a similar diet of liquids only. Stenosing esophagitis, with or without erosions and ulcerations, requires dietary management similar to that of peptic ulcer, with the added necessity of having the food finely divided to enable it to pass the narrowed area. Antacids also should be administered as needed for the control of pain, heartburn, gas, and sour eructa-

tions When the narrowing is not too marked gruels cereals strained vegetables and ground meat may be tolerated Each patient s problem must be considered individually and it should be remembered that patients usually tolerate best the food and drink that they find palatable There are however certain obviously undesirable items such as alcohol and fried or highly seasoned foods

MEDICINAL THERAPY

Antacids are useful in the treatment of any inflammatory disease of the esophagus — especially esophagitis with or without erosions, ulcerations or stenosis Aluminum hydroxide is probably the best of the antacids it is sold in liquid or tablet form as Amphojel Creamalin, or Gelusil Sodium bicarbonate also is helpful but if its use is prolonged it contributes to the formation of kidney stones and the production of metabolic alkalosis

Antispasmodics are of value in the control of spasm So called cardiospasm or achalasia is often helped by the inhalation of an amyl nitrite pearl or by the use of nitroglycerin grains 1/200 placed dry under the tongue just before a meal X ray as well as clinical evidence supports the observation that barium as well as food may enter the stomach more readily under the relaxing effect of amyl nitrite or nitroglycerin There is frequently an element of spasm in inflammatory diseases of the esophagus and in such cases some symptomatic relief may be obtained by the use of nitroglycerin but treatment of the underlying disease must receive first consideration Atropine sulfate is an antispasmodic which is useful in pylorospasm but has little effect in esophageal spasm or achalasia Morphine sulfate relaxes the patient as a whole and thus contributes to the relief of esophageal spasm The barbiturates have a similar action but these cannot be used indiscriminately owing to the danger of drug addiction

Psychotherapy

All too frequently the physician who is confronted with a patient who has difficulty in swallowing is tempted to say that it is all caused by "nerves." He may not even order an x-ray examination and may give no treatment at all, or he may merely try to reassure the patient by suggesting that he "forget all about it." If, however, he does order an x-ray and finds that it reveals spasm, he may then content himself with psychotherapy. Unfortunately, if this procedure is followed, such conditions as cancer of the stomach, cancer of the esophagus, esophagitis, esophageal ulcer, and foreign body in the esophagus can be neglected. Of prime importance is an adequate x-ray examination. Esophagoscopy with biopsy is essential to complete the diagnostic study, and even then negative reports should not be taken seriously if symptoms persist and the underlying cause of the spasms has not been determined. Cytologic studies also should be made.

Are there any diseases of the esophagus in which psychotherapy is of paramount importance? Yes, in the rare case in which all organic disease has been excluded psychotherapy should be tried. Psychotherapy also may occasionally be used in achalasia, but only in conjunction with the usually accepted treatment by nitroglycerin and bouginage. Even then it is probably best not to send the patient to a psychiatrist but merely to suggest to him that since the large mercury-filled bougie passes readily into the stomach food also will pass. In other words, the physician who passes the bougie can himself provide at least reassurance and support. Patients with achalasia may eat well when alone but not in company. Therefore emotional strain must be avoided, but the services of a psychiatrist are not usually needed.

Bouginage

Bouginage is a very useful therapeutic procedure in the treatment of web, benign stenosis, lye stricture, inoperable carcinoma, and achalasia.

Types of bougie

There are four types of bougie

(1) *Flexible silk-woven tipped bougies* with a steel shaft (see Figure 9) are passed under direct vision through the esophagoscope These bougies are made in graduated sizes from #8 to #26 French the latter being the largest size that will pass through the standard full lumen 9 mm Jackson esophagoscope Larger sizes may be used through larger esophagoscopes but the 9 mm instrument is chiefly used

(2) *Flexible spiral tipped bougies* with graduated olives and a stiff whalebone staff (Fig 14) may be passed in the office or in the

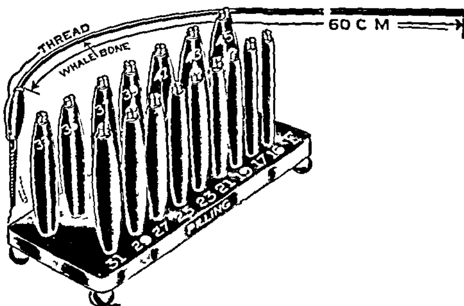


FIGURE 14 Esophageal dilators with whalebone staff flexible spiral tip and dilators of various sizes for use in benign stricture caustic stenosis and carcinoma of the esophagus

out patient department with a previously swallowed thread being used as a guide Any cooperative patient who can swallow his own saliva can swallow a #1 silk thread After about twenty feet of

thread have passed through the stomach into the intestine the thread *becomes firmly anchored* and may be pulled tight at the mouth to act as a guide for the passage of bougies This technic eliminates the danger of the bougie s entering a false passage or a diverticulum The only serious danger is that of dilating the esophagus too much at one sitting It is advisable not to pass more than three bougies at one session and to stop when there is resistance or bleeding

Directions for swallowing a thread to be used as a guide for bouginage are as follows

Obtain a large spool of #1 white silk thread from a surgical supply house Fasten the spool on the patient s clothing or in the upper breast pocket

Instruct the patient to take the end of the thread in his mouth and swallow it with water or any liquids or foods *It sometimes* helps to attach the thread to a gumdrop or a "lifesaver Do not put any lead shot on the end of the thread and do not tie knots anywhere in the thread except at the end if fastening it to a gum drop or 'lifesaver Instruct the patient to swallow the thread slowly — at a rate no faster than six inches per hour — otherwise it will become tangled in the esophagus Do not awaken the patient *at night for purposes of thread swallowing*

Cut the thread at the anus when it appears with the bowel movement Do not cut the thread between the spool and the mouth

Do not pull the thread at the mouth to see if it is anchored, it usually requires two *or three days for it to be firmly* anchored

For measurement mark the thread with one ink mark at five feet two ink marks at ten feet, three ink marks at fifteen feet and four ink marks at twenty feet

(3) *The Hurst rubber-covered mercury-filled bougie* (Fig 15) *is very useful in the treatment of achalasia* Because it is blunt and rounded this is the only type of bougie which may be passed blindly without danger The usual size is #50 French Most patients need no preliminary sedation or cocainization However a few hours previous fasting is advisable The patient should sit in a

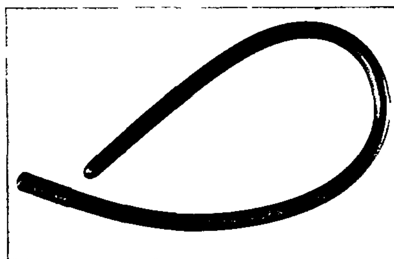


FIGURE 15 Hurst mercury filled bougie—very useful in achalasia.

straight chair head bent forward with chin on chest to relax the cricopharyngeus breathing through the mouth As the bougie passes the cardiac orifice there may be a slight delay and the patient may feel transient subxiphoid pain A modification of the Hurst bougie is the *Browne McHardy mercury bougie* with dilating bag attachment (Fig 16) By means of the dilating bag the cardiac orifice can be stretched to a diameter of 3 or 4 cm Accidents may happen if too great a dilatation is carried out

(4) *The retrograde bougie* In our experience retrograde bouginage except perhaps in the treatment of lye stricture in children is rarely indicated If peroral bouginage is unsuccessful in the adult it probably means total atresia in which case surgery is safer and hence preferable to the combination of peroral and retrograde bouginage Lye strictures in children are usually treated by retrograde bouginage Gastrostomy is performed for feeding a thread is swallowed and pulled out through the gastrostomy opening and the retrograde bougies are attached to the thread and pulled up through the stomach and esophagus and out the mouth It is probable that small children are more benefited nutritionally and psychologically by this method than by peroral bouginage

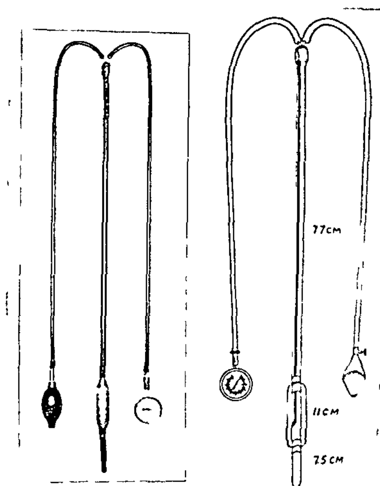


Figure 16 Browne McHardy mercury bougie with dilating bag attachment (Courtesy of *Journal of American Medical Association* 113 1963, Nov 1939)

Bougienage in benign peptic stenosis

Benign peptic stenosis seems to be chiefly due to the regurgitation of acid peptic gastric secretions into the lower esophagus resulting in esophagitis with or without erosion and superficial ulcerations, and peptic ulcer It is often accompanied by hiatus hernia and is frequently associated with duodenal ulcer Unless the stenosis has progressed so far as to narrow the lumen greatly and cause irreversible fibrosis good results can be obtained by bougienage

The first bouginage is performed at the time of the first esophagoscopy at which time the flexible-tipped bougies of varying sizes are passed under direct vision through the esophagoscope. A biopsy specimen should always be obtained at the same time to exclude carcinoma. Bouginage should precede biopsy so that the danger of perforation may be minimized. In experienced hands the risk of perforation from bouginage and biopsy is negligible. Follow-up studies were carried out in a series of 100 cases reported by Benedict and Gillespie (1). Thirty-four of the patients were seen at least one year after the last treatment: 17 were able to swallow all food normally, 15 were able to swallow liquids and soft foods but had some difficulty with solid food, and 2 were unchanged. Others are still under treatment and are progressing so well that surgery has not been seriously contemplated. Some of the poor risk and elderly patients are content to subsist on liquids and strained or ground foods rather than undergo an operation.

Bouginage in lye stricture

In children retrograde bouginage as outlined above is usually the treatment of choice. In adults the first bouginage is performed under direct vision at the time of esophagoscopy. Since carcinoma occasionally develops in lye stricture, it is well to make one direct inspection. Inspection, however, is limited to the upper stenosis since the esophagoscope will not usually pass beyond the area of narrowing. The best type of bouginage, therefore, is the peroral method with a previously swallowed thread being used as a guide. Retrograde bouginage necessitates gastrostomy, but this is very seldom necessary and should be avoided if possible. The response to peroral bouginage varies according to the degree of stenosis. When there are several strictures and much esophagitis the prognosis is poor. Clinical improvement is usually more marked than x-ray improvement. If by means of repeated bouginage a lumen of 11 or 12 mm (= 35 to = 37 French bougie) can be maintained, it should be possible for the patient to eat normally if all food is well chewed.

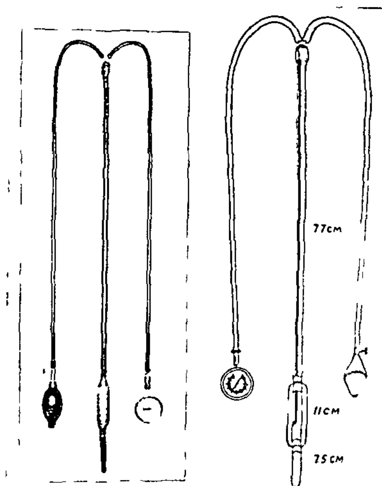


FIGURE 16 Browne McHardy mercury bougie with dilating bag attachment (Courtesy of *Journal of American Medical Association* 113 1963, Nov 1939)

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Surgery must be considered if the patient is a good risk and if the response to bouginage is unsatisfactory

Bouginage in carcinoma

Carcinoma of the esophagus should be resected in a good risk patient Even if the result is only palliative, resection is probably worth while in most cases Unfortunately, however, many cases of carcinoma are inoperable In such cases a combination of bouginage and x-ray treatment may result in considerable palliation and will obviate a gastrostomy It is well to have patients swallow a thread when x-ray treatment is started, otherwise radiation fibrosis may obliterate the lumen When there is no lumen at all a dimple may be visualized by esophagoscopy, and bouginage under direct vision at that point may result in the opening of a satisfactory lumen If a thread is kept in place throughout the period of x ray treatment there is no danger of obliteration of the lumen Patients with inoperable carcinoma of the esophagus prefer to eat by mouth even if the diet is quite restricted, rather than be fed by gastrostomy The results of peroral bouginage in carcinoma of the esophagus are quite unpredictable In general the best results are obtained in the radiosensitive tumors On rare occasions there are brilliant results with complete eradication of the local malignancy

Bouginage in achalasia

The first bouginage in achalasia is often performed under direct vision through the esophagoscope However, the largest bougie that can be passed through the 9 mm esophagoscope is the #26 and this is not large enough to stretch the cardiac orifice adequately hence little is accomplished unless the esophagoscope itself *is passed into the stomach* For this purpose the 53 cm esophagoscope is best because of its length Under local anesthesia and sometimes even under general anesthesia it may be difficult to find

and pass the cardiac orifice Usually more can be accomplished by the use of the Hurst mercury filled bougie Sometimes when the diagnosis seems obvious on the basis of the patient s history and x-ray examination and there seems to be no possibility of any complicating factor, esophagoscopy may be omitted If however there is the slightest doubt about the diagnosis esophagoscopy must be performed Later bouginage with the Hurst bougies yields good results in most cases even one treatment sometimes results in miraculous improvement In other cases treatments must be given at periodic intervals — monthly weekly or even daily If frequent treatments are necessary some patients can be taught to pass the bougie themselves In intractable cases greater dilatation is achieved by using the Browne McHardy bougie although even here the results are unpredictable When medical treatment and bouginage fail surgery is indicated

Complications of bouginage

If properly performed in a good-risk patient by a skillful and experienced physician bouginage involves almost no risk Blind bouginage (except with the mercury bougie) should never be carried out because of the danger of perforation into the mediastinum when the instrument has entered a false passage or diverticulum Fluoroscopic guidance does not ensure safety in blind bouginage since the fluoroscope cannot help to guide the bougie away from a false passage or diverticulum An unsuspected bolus of meat or some other foreign body might be encountered during the process and this might lead to perforation mediastinitis and death Bouginage is most successful and least dangerous when the patient is well hydrated it should not be attempted in a dehydrated person

Bouginage under direct vision through the esophagoscope is a safe method in a good risk patient The endoscopist can see the lumen at its upper end and guide the bougie directly into it

Thereafter as long as the esophagoscope is held in the axis of the lumen the bougie will enter and dilate the stenosed sector Occasionally the lumen seems to be completely obliterated, or there may be only a dimple to suggest its location Under these circumstances gentle probing with a small bougie may reveal the lumen and dilatations can be carried out Progression to larger bougies may continue only as long as there is very little resistance and very little bleeding In our experience only one perforation has occurred when this method was used, and that was in a bad risk elderly morphine addict who had successfully undergone bougienage on 21 occasions but sustained a perforation during the twenty second esophagoscopy and bougienage He showed no reaction to the perforation and no resistance to the dilatation He died on the following day On rare occasions bleeding may result from the irritation of an esophageal ulcer by bougienage, in one such instance the bleeding was severe enough to necessitate transfusion Esophageal ulcers, like other ulcers, may bleed spontaneously, but when bleeding occurs immediately after bougienage the endoscopist is considered responsible

Bougienage using a previously swallowed thread as a guide is quite safe if properly performed Among all the cases of benign and malignant stenosis with which we are familiar only one perforation has occurred the reason being that a member of the house staff attempted too-rapid dilatation, passing eight bougies at one sitting and progressing too soon to an excessively large bougie Here again hemorrhage from an ulcer or from a severely eroded area of esophagitis may be initiated by bougienage

Bougienage using the Hurst mercury-filled bougie In our experience no complications of any sort have occurred with the use of this soft blunt, rubber-covered bougie If it fails to pass the cardia it simply stops completely or in megaesophagus curls up benignly in the dilated lower portion of the esophagus With the Browne-McHardy bougie perforation can be caused by too great a pressure on the cardia Fortunately, however this has not occurred in our experience

Retrograde bougmage is a very safe method of dilating the esophagus There is little risk of perforation or hemorrhage When the thread is pulled upwards care must be taken not to cut the tongue

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SURGICAL PRINCIPLES

7

PREOPERATIVE AND POSTOPERATIVE CARE

Surgery of the esophagus is indicated either primarily, as in tumors, *diverticulum, rupture and fistula, or secondarily after* failure of conservative treatment, as in achalasia, web, stricture and ulcer.

The preoperative preparation of the patient who is to undergo esophageal surgery is of paramount importance. The procedure to be employed depends on the surgeon's evaluation of the patient as a surgical risk on the basis of general health, age and prognosis.

Patients with esophageal disease are frequently malnourished. Attempts should be made to improve their general nutrition and hydration by means of intravenous alimentation, giving high caloric liquids by mouth and in selected cases gastrostomy. However, *time and effort should not be wasted in trying to achieve an* ideal nutritional status, for such attempts are usually futile, time consuming and not metabolically sound.

Surgery which is to include mediastinal dissection should be preceded by an electrocardiogram. *Evidence of cardiac irritability* in the tracing indicates that quinidine sulfate should be given both before and after operation to reduce the hazard of cardiac arrhythmia. It is advisable to give the patient a test dose of quini-

dine sulfate before operating to rule out sensitivity to the drug

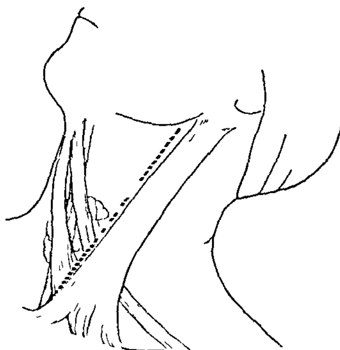
Chemotherapy is a valuable adjunct in esophageal surgery — particularly for the patient whose esophagus is to be opened in the course of the procedure We have favored parenteral administration of penicillin and streptomycin since we believe that these agents have a low incidence of side effects as compared with the newer wide spectrum antibiotics The patient receives these drugs just as he is leaving for the operating room so that a therapeutic blood level may be present at the time of potential contamination Chemotherapy should be continued after operation for a period of 5 to 7 days in adequate doses (*e g* , 300 000 units of penicillin and 0 5 gm of streptomycin every 8 hours) and then omitted At this time if residual sepsis is present it is better for it to localize and thus be identified so that it may be properly treated than for it to be masked by prolonged administration of antibiotics

Some physicians believe that the oral administration of anti biotics (such as 0 25 gm of streptomycin in a glass of water t i d) may appreciably reduce the gastroesophageal flora and hence the incidence of sepsis in resective surgery of the esophagus We are not convinced of its effectiveness

It is advantageous to have a nasogastric tube in place during operations on the esophagus Not only does it serve a useful purpose in decompressing the stomach but in addition its presence within the esophageal lumen aids the tactile finger in the course of dissection In accordance with our belief that it is poor surgical practice to leave any foreign body on an anastomotic suture line we remove the tube at the completion of the operation or within 4 to 48 hours thereafter Prolonged utilization of a tube is probably an important factor in the development of acute esophagitis The use of a nasogastric tube is contraindicated in the presence of bleeding esophageal varices

General Surgical Incisions and Approaches

To avoid repetition in later chapters the principal surgical approaches to the esophagus will be described here



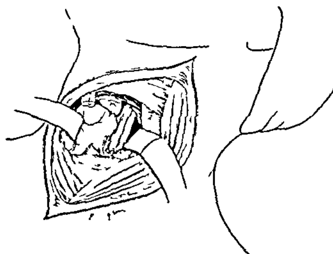


FIGURE 17 Approach to the cervical portion of the esophagus Incision along the anterior margin of the sternocleidomastoid muscle

Cervical portion of the esophagus

The cervical portion of the esophagus which lies slightly to the left of the midline is best approached through the left side of the

neck With the patient's head turned to the right a long, straight incision is made along the anterior border of the sternocleidomastoid (Fig 17) The left lobe of the thyroid gland and the sternohyoid and sternothyroid muscles are retracted medially The omohyoid is identified and retracted downwards or divided The ansa hypoglossi may be divided without harm The carotid sheath is retracted laterally The esophagus can easily be reached beneath the trachea

This approach allows excellent access to the cervical esophagus it is utilized in excision of diverticula drainage of the superior mediastinum and esophageal anastomosis at this level

Upper thoracic portion of the esophagus

The upper thoracic portion of the esophagus may be approached through either the right or the left side of the chest The choice is dictated by both the surgeon s personal preference and the problem at hand Because of its curvature to the right the accessibility of the azygos vein and the absence of the aortic arch the upper esophagus is usually most easily dealt with through a high right thoracic incision (Fig 18)

Resection of the fifth rib or in younger patients incision

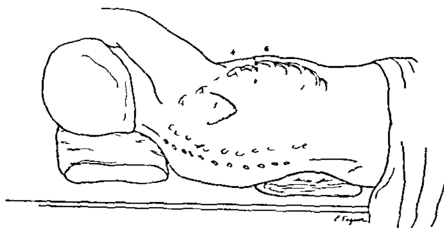


FIGURE 18 Approach to the upper thoracic portion of the esophagus Incision over the right fifth rib

through the fifth intercostal space provides excellent exposure of the upper thoracic segment of the esophagus Should mobilization of intra-abdominal viscera such as stomach or small or large bowel be necessary a separate abdominal incision is made

Separate *abdominal and thoracic incisions frequently* entail shifting positions changing gowns and linen, and increased risk of contamination

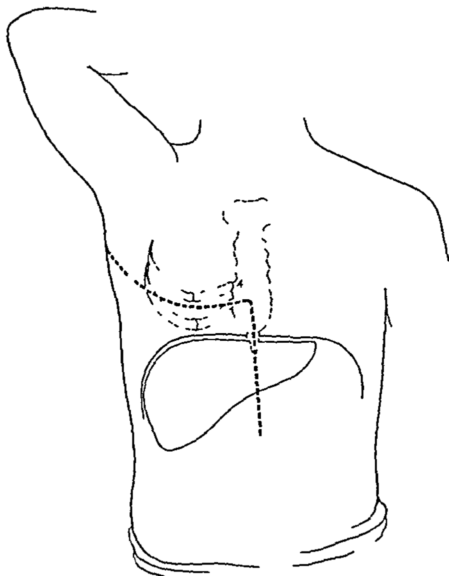


FIGURE 19 The thoracosternoabdominal approach of Moore

There are two alternatives The first is a high left thoracic approach with incision of the diaphragm or a left thoracoabdominal incision (the latter is rarely necessary) The second which has been described by Moore (2), is a thoracosternoabdominal incision with the patient supine The incision is carried down vertically over the sternum splitting the latter in its lower third, and is then continued as an abdominal incision (Fig 19) We have found this a useful approach in selected cases Lawrence (1) has recently described a right thoracoabdominal approach to esophageal resection (Fig 20)

Lower thoracic portion of the esophagus

The best approach to the distal third of the esophagus is through the left lower chest at the level of the seventh or eighth rib (Fig 21) A lower approach may place the surgeon at a disadvantage since the dome of the diaphragm will lie between him and the operative site

This incision can easily be extended into a thoracoabdominal incision when desired In cases of carcinoma it may be desirable to explore the abdomen for metastases before opening the chest In such instances the abdominal portion of such a thoracoabdominal incision is made first

Pyloroplasty

As more experience is gained in esophageal surgery the role of gastric retention and of acid peptic regurgitation in producing post operative complications is becoming more and more apparent The importance of ensuring an adequate and efficient channel for gastric emptying after any esophageal procedure cannot be overemphasized The frequent necessity of vagotomy the high incidence of associated pyloro duodenal disease and the distortion attendant on gastric mobilization make it imperative for the surgeon to judge whether or not his patient will have postoperative

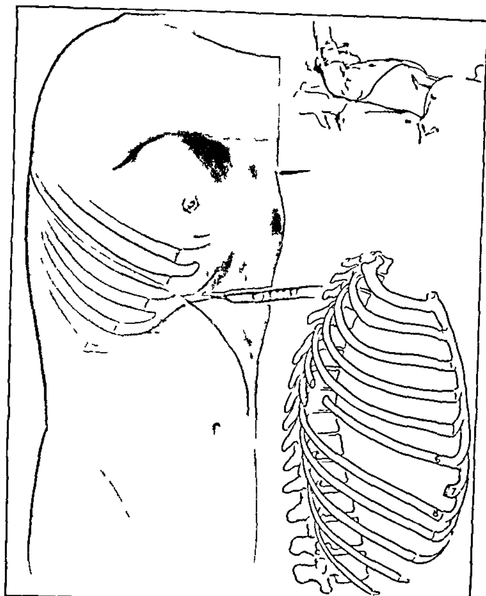


FIGURE 10 Right sided thoracoabdominal incision carried from the posterior axillary line to the left paraumbilical area (By permission of the *Journal of Surgery Gynecology and Obstetrics*)

difficulty with gastric emptying This is equally important whether the procedure contemplated be for hiatus hernia achalasia stricture or carcinoma If difficulty is anticipated a concomitant pyloroplasty may be indicated We have found the Heineke-Mikulicz procedure simple and effective In cases of esophago

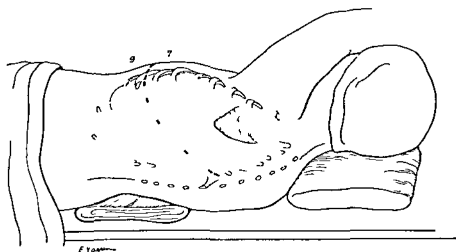


FIGURE 21 Conventional posterolateral approach to the lower esophagus through the seventh or eighth rib

gastrostomy in which the stomach is to be transplanted into the chest and the blood supply to this organ depends solely on the right gastric and gastroepiploic arteries the surgeon must bear in mind the possibility of compromising this blood supply in executing such a pyloroplasty

Gastrostomy

The indications for this procedure are rapidly diminishing With rare exceptions its value lies only in improving the general nutrition of patients with congenital esophageal atresia or extensive strictures of the esophagus (e g lye stricture) In these cases the gastrostomy is repaired as soon as it has served its purpose

Gastrostomy provides no palliation of esophageal cancer and may merely prolong the patient s suffering The Stamm gastrostomy is the simplest safest and most effective

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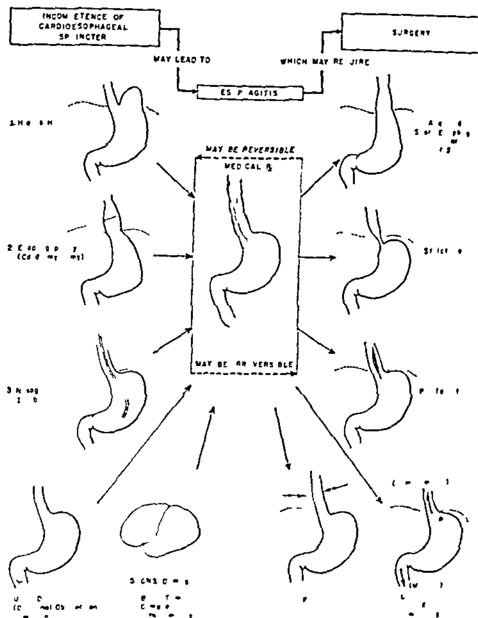
Esophagitis

8

Esophagitis is probably the commonest disease of the esophagus (Fig 2) Nonspecific esophagitis may be of bacterial traumatic or neurogenic origin Several factors may combine to produce esophagitis for example the bacteria normally present in the lumen may gain entry into mucosa irritated by constant bathing with acid gastric secretions from a hiatus hernia

Etiology

In 1938 Selye (8) produced in rats a condition which he termed peptic hemorrhagic esophagitis by placing a ligature around the pyloric end of the stomach and thus causing a marked accumulation of gastric juice with regurgitation into the esophagus Bacteria normally present in the esophageal lumen include *Staphylococcus Streptococcus Pneumococcus B coli*, and Friedlander s bacillus Carious teeth and diseased tonsils probably contribute a large number of organisms to the esophagus Normally the combination of intact esophageal mucosa and the natural bodily resistance prevents bacteria from producing any inflammatory reaction Any trauma to the esophagus immediately enables bacteria to produce esophagitis Excessive intake of alcohol and the long continued use of



FIGURE

highly seasoned or very hot food may traumatize the mucosa and lead to esophagitis Caustics produce burns of the esophagus leading to esophagitis and stenosis Foreign bodies in the esophagus unless promptly and skillfully removed are very likely to produce trauma and infection Inlying Levin Harris or Miller Abbott

tubes may irritate the esophageal mucosa and contribute to the development of esophagitis Extrinsic pressure on the esophagus caused by malignant mediastinal or bronchogenic tumors or malignant or tuberculous lymph glands may cause marked deviation of the organ, with consequent delay in the passage of food and then irritation and esophagitis Some esophagitis is always found in long standing achalasia because this condition is characterized by esophageal obstruction stagnation of food and secretions, and multiplication of bacteria Persistent vomiting such as may occur after operation and in gallbladder disease and pyloric stenosis almost certainly produces trauma to the lower esophagus through the action of gastric juice if the body in general and the esophageal mucosa in particular have little resistance esophagitis is very likely to occur Ill advised surgical procedures such as esophagoplasty may be complicated by the development of severe esophagitis (This problem will be considered in subsequent chapters)

There is much debate regarding the importance of reflux gastric juice in the development of esophagitis We believe that this factor is of very great importance In 100 cases of peptic stenosis of the esophagus recently reported by Benedict and Gillespie (4 5) 87 patients had a hiatus hernia which apparently favors the presence of reflux gastric juice and the development of esophagitis However 13 per cent of these patients had no demonstrable hernia but some had suffered from gallbladder disease and others had had vomiting owing to peptic ulcer of the duodenum or stomach pyloric stenosis alcoholism or pregnancy On the other hand it must be admitted that hiatus hernia occurs in many persons without causing symptoms and apparently without producing esophagitis Furthermore it is known that esophagitis may be found in the absence of hiatus hernia Patients in the latter group however had always had vomiting or regurgitation of gastric juice

What part does heterotopic gastric mucosa play in the development of esophagitis So far as we can determine on the basis of all our cases and of a review of the literature the gastric mucosa which is obtained by esophagoscopic biopsy or surgical resection

duration. The patient's past history revealed 27 years of heartburn with one sudden episode of hematemesis. Fourteen years before she had been put on a diet because of "scarred stomach." Ten years later she had had burning pain in her stomach shooting through to her back. In the 6 months before admission to the hospital she had lost [illegible] pounds. Gastric analysis revealed no free hydrochloric acid after the administration of alcohol and of histamine. X-ray examination demonstrated extensive scirrhous carcinoma of the stomach. Transthoracic total gastrectomy with esophagoduodenostomy was performed by Dr Soutter. The pathologic diagnosis was scirrhous carcinoma. The one lymph node submitted was negative. The patient did well after operation except that the substernal burning continued. Three months after operation a G I series was negative.

For a year and a half after the operation she continued to have fullness, nausea, regurgitation, epigastric pain radiating through to her back, anorexia and loss of weight. Recurrent carcinoma was suspected. A G I series was again negative. Esophagoscopy, however, disclosed a bleeding, soft, polypoid, nodular lesion which seemed to involve the entire circumference of the esophagus and to extend from the upper to the mid portion of the esophagus without notably obstructing the lumen. A satisfactory biopsy specimen was obtained. The findings were considered to be due to recurrent carcinoma. The pathologic diagnosis, however, was severe acute and chronic inflammation with granulation tissue. One year later the patient was eating well and had gained weight. She still had burning pain and biliousness. A later follow up disclosed that she continued to have difficulty in swallowing for the rest of her life. She died about 2 years after the esophagoscopy of carcinomatosis which almost certainly arose from the scirrhous carcinoma of the stomach.

Comment. This unfortunate woman seems to have had burning pain for nearly half her life. We cannot be sure how much of it was due to esophagitis. There are two features of special interest in this case: (1) Very severe esophagitis may not be visible at x-ray examination, and (2) very severe esophagitis may be so polypoid in appearance that it is mistaken by the esophagoscopist for carcinoma.

Treatment

The treatment of simple uncomplicated esophagitis is dietary, it includes avoiding alcohol and highly seasoned foods and living on a bland diet with frequent feedings However in severe esophagitis it may be advisable to give nothing by mouth and to use parenteral feedings Bland liquids are generally tolerated as soon as nausea and vomiting have subsided Elevation of the head of the bed may be helpful Commercially prepared baby foods, containing strained vegetables and ground meat, are useful Antacids are of aid in that they provide symptomatic relief The use of tobacco in any form should be avoided

Relation of Esophagitis to Peptic Ulcer of the Esophagus

Can esophagitis progress to peptic ulceration We believe that it can since many of our patients gave an early history of dysphagia and heartburn with no evidence of ulceration but years later were found at operation to have a true peptic ulcer We consider the following case a probable instance of esophagitis progressing to ulceration

M C a 30-year old man entered the hospital with a 3½ year history of dysphagia X ray examination on three occasions showed (1) gastric hernia and congenitally short esophagus (Army report) (2) duodenal ulcer and hiatus hernia and (3) normal esophagus with 8 cm hiatus hernia On the fourth examination made four years after the first x ray the esophagus was found to be slightly narrowed for 3 cm above the hernia and there was a questionable esophageal ulcer Esophagoscopy first showed inflammation and narrowing of the esophagus and later revealed very little narrowing slight esophagitis and gastritis in the hiatus hernia Biopsy confirmed the presence of slight chronic inflammation of gastric mucosa One year after the patient was first seen by one of us (I BB) and four years after the onset of symptoms he required surgery because of moderately severe hemorrhage and the failure of medical treatment and bouginage Transthoracic par

tial esophagectomy and gastrectomy was performed by Dr R H Sweet (Dec 1947) the pathologic diagnosis being peptic ulcer of the esophagus active with fibrosis and stricture Five months later this patient was very much improved eating freely of a liberal diet but still having some heartburn and substernal discomfort

Comment We now wonder (1) whether the so called esophageal ulcer was really *of gastric origin (on reviewing the slides the* pathologist is not certain and (2) whether the resection (a) re*moved the entire area of esophagitis and* (b) *removed* enough of the stomach to reduce the acidity Since some subsequent resections have been more extensive, in that more esophagus and more stomach were removed, the results have been symptomatically better

Relation of Esophagitis to Carcinoma

On the theory that chronic irritation may lead to carcinoma one should suspect that carcinoma might arise in areas of severe long standing esophagitis A few years ago one of us (I BB) (6) reported two cases of carcinoma of the esophagus, of which one had apparently developed in a lye stricture and the other in a so called congenitally short esophagus A third case has come to our attention in which carcinoma apparently developed in an area of esophagitis of 23 years duration

M D a 70 year old man first entered the hospital in 1944 with a complaint of substernal pain and dysphagia of 23 years duration A *duodenal ulcer had been suspected at an x ray* examination made in 1910 In 1923 an esophagoscopy was performed at another hospital this revealed a gross appearance said to be characteristic of carcinoma The pathologic diagnosis however was chronic inflammation During the next 21 years the patient had episodes of very severe pain melena and hematemesis for which he was treated by various excellent gastroenterologists and esophagologists in several large cities in *the United States and Europe receiving reports* of hiatus hernia and esophageal ulcers As recently as 1943 an eminent esophagologist had noted hiatus hernia stenosis and superficial ulceration but nothing was noted that suggested cancer and a biopsy specimen was not obtained

The first esophagoscopy to be done at this hospital was performed by one of us (EBB) in 1944. The gross appearance was again found to be consistent with acute esophagitis and benign stenosis. This time, however, a biopsy was taken which revealed adenocarcinoma. (See Figure 31.)

Comment. We consider that this case not only emphasizes the importance of biopsy but also illustrates the possibility that carcinoma may arise in long standing esophagitis or gastritis in hiatus hernia.

Agranulocytic Esophagitis

A case of marked esophageal stenosis following agranulocytosis due to sulfonamide therapy has been reported by Bryan (7). This patient had been given 1 gm. of sulfadiazine every 3 hours for 8 days because of sore throat. The medication had been administered by another physician but the patient had become steadily worse and a peritonsillar abscess had developed; this had been opened and drained. Ulcerations of the buccal mucosa gradually became fulminating. The white-cell count fell to 1,200. The patient's temperature rose to 106° F. Sulfadiazine had been stopped on admission to an outside hospital and intramuscular administration of penicillin had been started. After a very stormy 2 weeks course the patient began to improve; he was finally discharged home and began to gain weight. However, on leaving that hospital the patient had had some dysphagia and this gradually progressed for the next 5 months until he could swallow only fluids or thin gruels. X-ray examination revealed a long funnel like narrowing that began at the level of the carina. On esophagoscopy at another outside hospital the narrowing was visualized and the mucosa was seen to be pale and lusterless. Bouginage was carried out on several occasions with improvement and gain in weight. After 2 or 3 weeks, however, dysphagia returned. The patient was admitted to this hospital where esophagectomy was performed by Dr. R. H. Sweet with complete relief of the dysphagia.

Idiopathic Ulcerative Esophagitis

A very unusual case has been reported by Achenbach Lynch and Dwight (1) in which the ulcerative disease was primary in and confined to the esophagus Associated with this primary disease were migratory arthritis erythema nodosum, dermatitis gangrenosa, and multiple perianal abscesses and fistulas In many ways this condition resembled ulcerative colitis

Esophagectomy apparently cured the associated arthritis skin lesions and perianal condition

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Hiatus Hernia

9

Hiatus hernia results from displacement of the stomach and the distal portion of the esophagus through the esophageal hiatus into the thorax It is a form of diaphragmatic hernia Diaphragmatic hernia not involving the esophagus will not be considered here

Incidence

Over a thirteen year period in a city of 30 000, Mobley and Christensen (26) encountered 153 patients with esophageal hiatus hernia On the basis of their experience they projected the incidence of this condition in the United States as 5 per 1000 In 3448 x ray examinations of the gastrointestinal tract for digestive symptoms hiatus hernia was found to be the second commonest abnormality (7) Duodenal ulcer was present in 20 4 per cent of these patients A hiatus hernia was observed in 8 9 per cent

Herniation may occur at any age but is most common in the fifth and sixth decades Men and women are equally affected

The increased abdominal pressure in the latter part of pregnancy results in a high incidence of transient hiatus hernia with heartburn as a symptom (20 34) Treatment is symptomatic The condition

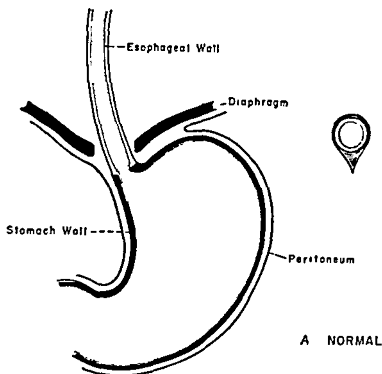
is usually self-limited and has no effect on the outcome of the pregnancy. Its etiology is unknown.

Types

Esophageal hiatus hernia may be classified into 3 groups: (1) Hiatal or sliding, (2) parahiatal or rolling, and (3) short esophagus with thoracic stomach (possibly congenital).

True hiatal or sliding hernia is the type most frequently encountered; it represents the majority of cases seen by the surgeon. In this group the cardia passes into the thorax through an enlarged hiatus. It is a true sliding hernia comparable to sliding hernia of the cecum. The stomach forms one of the walls of the hernial sac. The peritoneal portion of the sac lies anteriorly, extending around each side (Plate VIIB). Such sliding hernias are associated with an esophagus of normal or less than normal length. Esophageal shortening is probably acquired as a result of cicatricial contracture secondary to esophagitis and ulceration. It is impossible to predict before operation whether or not significant organic shortening of the esophagus will be found (36). X-ray studies are particularly unreliable in such cases (39). This group of sliding hernias can accordingly be subdivided into those with an esophagus of normal length and an esophagus with acquired shortening. This distinction will assume some importance in our discussion of congenital short esophagus.

Parahiatal hernia occurs less commonly than sliding hernia (5 to 15 per cent) (37, 41). The cardia remains in its normal position surrounded by a loop of diaphragmatic fibers which lie between it and the hernial sac (Plate VIIC). (It is accordingly not a true hiatus hernia, hence the term "parahiatal" rather than "paraesophageal.") The hiatus remains normal in size, whereas the hernial aperture may become quite large. As it increases in size the stomach rolls into it and assumes an inverted position popularly termed "upside down stomach." Fleischner (17) contends that the indentation between esophagus and herniated gastric pouch seen on



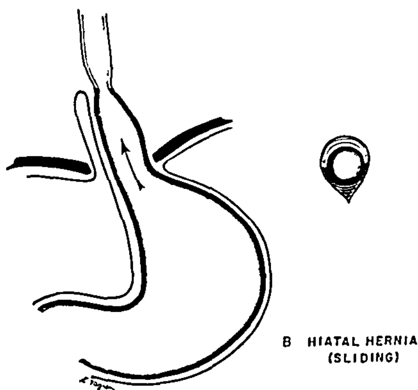


PLATE VII Diagram to illustrate the anatomical relations in hiatus hernia The esophagus is shown in yellow the stomach in red and the peritoneum in green (A) Normal relations A cross section through the hiatus shows only esophagus. (B) In sliding hernia the stomach is displaced upwards and the peritoneal sac is anterior

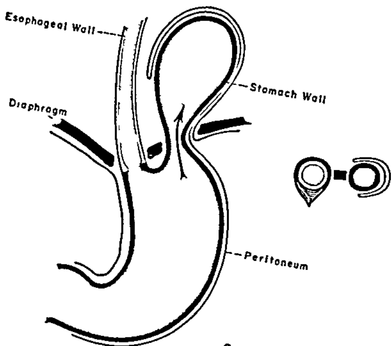




PLATE VII (*continued*) (C) In parahiatal hernia the cardia remains in its normal position (D) In a true congenitally short esophagus the cardia has failed to descend to its normal position and there is no hernial sac

barium swallow represents the persistent incisura cardiaca rather than a fasciculus of diaphragmatic muscle. The combination of hiatal and parahiatal hernia may occur.

True congenitally short esophagus with thoracic stomach is rare. Most cases so classified are probably not congenital abnormalities but represent secondary shortening of the esophagus subsequent to esophagitis induced by reflux of gastric contents (Plate VIII). In 87 patients with x-ray findings suggesting congenitally short esophagus Sweet (38) found only 4 at operation—and we doubt that even these represent true congenital shortening. In a review of 220 patients with hiatus hernia and short esophagus by Olsen and Harrington (29) only 9 (4 per cent) were considered to have true congenitally short esophagi. Congenital short esophagus with thoracic stomach is a true developmental abnormality. The diaphragmatic septum transversum originates from the cervical mesoderm and during embryonic development descends caudally with the phrenic nerve. During its development the stomach descends to overtake and pass the septum transversum. The stomach enters the abdomen before the lateral diaphragmatic components fuse with the septum transversum. If gastric descent is slow the stomach may become trapped in the thorax by the diaphragm and a fixed hernia with short esophagus results. It should be noted that no peritoneal hernial sac results from this asynchronous fusion: the stomach is partially in the thorax and covered by mediastinal pleura but not by peritoneum (6). Another distinctive feature in this rare group is a direct segmental arterial blood supply from the intrathoracic aorta to the intrathoracic stomach (1). If subsequent cicatricial shortening ensues in such a situation a sliding type of hernia could occur and a partial peritoneal sac could develop in its usual anterior position. Such a combination of developmental and acquired defects would cause the patient to have a thoracic stomach and a markedly shortened esophagus.

The term "congenital short esophagus" is a poor one since acquired shortening could occur before birth and in this sense be congenital.

We would recommend the term 'developmentally short esophagus' for those cases in which the short esophagus and thoracic stomach result from developmental errors. In these cases no hernial sac will be found. It is not a true herniation of a once normally situated organ. For the more common acquired short esophagus we suggest the term "hiatus hernia with shortened esophagus."

Potter (3[illegible]) has never observed a congenitally short esophagus with thoracic stomach in her experience at the Chicago Lying-in Hospital. Even with absence of the major portion of the diaphragm she found the esophagus to be of normal length. Husfeldt *et al* ([illegible]) feel that a congenital short esophagus does not exist or is extremely rare. Uebermuth (40) feels the condition should not be considered a hernia, the two affections having in common only gastroesophageal reflux and the resulting esophagitis. D'Aste (13) feel that the congenitally short esophagus with a fixed stomach and no hernial sac should not even be grouped with hiatus hernias.

From a practical point of view it is probably better to accept the fact that the symptomatic short esophagus associated with hiatus hernia is acquired and inflammatory rather than of developmental origin.

Symptoms

Hiatus hernia is frequently asymptomatic. Symptoms depend on the type of hernia. There is little relation between the size of the hernia and the distress resulting therefrom.

The symptoms in sliding hernia are usually esophageal in origin. The competence of the esophagogastric junction is due to the oblique insertion of the esophagus into the stomach, the intrinsic sling fibers of the stomach, the musculature of the right crus, and the physiologic cardiac sphincter. The upward displacement of the esophagogastric junction destroys the efficiency of these valvular mechanisms and the subsequent regurgitation of acid peptic contents in the esophagus results in symptomatic esophagitis.

The commonest symptom of sliding hiatus hernia is a burning

retrosternal or epigastric pain This is worse when the patient is lying down and is exacerbated by stooping or straining It is relieved by sitting upright and taking water or antacids Dysphagia is the next most common complaint

The symptoms of hiatus hernia which include radiation of pain down the left arm occasionally simulate those of coronary heart disease The electrocardiogram is the most reliable special method of distinguishing cardiac from esophageal pain

In parahiatal hernia the normally situated cardia is not incompetent and esophagitis does not result Symptoms are caused by the presence of the stomach or other viscera in the hernial sac they consist of postprandial fullness, shortness of breath and acute bouts of epigastric pain

Vomiting is the principal and invariable symptom in children with short esophagus and thoracic stomach (9) Additional symptoms may be those of reflux esophagitis

Heartburn hematemesis and melena may occur in both hiatal and parahiatal hernias The bleeding originates in the esophagus or in a segment of inflamed or ulcerated stomach trapped in the hernia Patients with hiatus hernia have a relatively high incidence of associated cholecystitis and diverticulosis coli This group of diseases is known as Saint s Triad (27)

Complications

Blood loss is the commonest complication and approximately 25 per cent of patients with sliding hernia have secondary anemia or a history of hemorrhage Clerf *et al* (10) found erosions of the gastric mucosa at gastroscopy in about 33 per cent of their patients Complete obstruction may develop The incarcerated viscera may become gangrenous and even perforation may occur (1 -3)

Diagnosis

Fluoroscopic examination with the patient in Trendelenburg position is the most reliable diagnostic maneuver It should be em

phasized that such examination may occasionally yield normal findings in patients with hiatus hernia and should be repeated when the clinical picture warrants it In a series of 115 patients with hiatus hernia the first x-ray examination was reported as negative in 63 (26) It should be re-emphasized that the roentgenologic diagnosis of shortened esophagus is unreliable

Esophagoscopy demonstrates normal or inflamed gastric mucosa in the hernia *Erosions and hemorrhages may be seen Since benign* stricture of the esophagus is often found in association with hiatus hernia (2, 3 4), the esophagoscopist is called upon to observe and treat such strictures and to distinguish them from other causes of esophageal obstruction He is also called upon in cases of hemorrhage in order to determine whether the bleeding is originating in the area of gastritis in the herniated portion of the stomach or in a possible esophageal ulcer Esophagitis is always present when there is any evidence of stenosis and is frequently present when there has been much regurgitation or vomiting Reddening of the mucosa of the lower end of the esophagus is a common finding There may be edema and narrowing and easy bleeding of the mucous membrane Sometimes in large hernias without stenosis the herniated portion is better inspected by means of the flexible gastroscope We have seen one case of large incarcerated hiatus hernia in which the diagnosis was made not by x-ray examination (recumbent position only) but by esophagoscopy, which showed gastric folds Subsequent x ray examination with the patient erect demonstrated the hernia (See Plate VI #5)

Benign stricture of the esophagus in association with *hiatus hernia* can frequently be handled satisfactorily by esophagoscopy and bouginage Incarceration is usually an indication *for surgery* but occasionally esophageal obstruction due to incarceration may be *considerably relieved by esophagoscopy and bouginage* Although such relief may be only partial the latter treatment should be *considered for the poor-risk patient* or as a preparation for surgery

TREATMENT

If the hernia is small and asymptomatic no treatment is needed. Asymptomatic patients should be followed closely, however. Sprafka *et al* (35) followed a group of such patients for as long as six years and found that 20 per cent of the small hernias progressed to large ones and complications developed in 30 per cent of the patients with large hernias.

Medical treatment

If symptoms are minimal, conservative management is indicated. A great many of such patients are overweight and the patient should be made to reduce. Constricting abdominal garments should be discarded and bending and lifting should be avoided. An ulcer regimen of bland diet, antacids, antispasmodics and sedatives is often helpful. If conservative measures are inadequate and symptoms of cardiac incompetence and esophagitis persist, surgery should be carried out.

Surgical treatment

The indications for surgery may be summarized as follows: (1) Intractability of symptoms; (2) massive or repeated hemorrhage with anemia; (3) stenosis not amenable to bouginage and medical management; (4) strangulation or obstruction; and (5) perforation.

The results of surgical treatment of hiatus hernia depend of course on the selection of cases and may be variable. Assuming that the patients are carefully selected and properly operated the results are usually gratifying. In a series of 130 surgically treated cases Sweet (37) had only a 3 per cent recurrence. On the other hand Gertz *et al* (19) followed 45 cases over several years and found only 5 per cent clinically and radiologically cured after operation.

Surgical repair of hiatus hernia may be carried out through either the abdominal or the thoracic approach The most important factors in the choice of approach are the experience and the personal preference of the surgeon

The objectives in the repair of hiatus hernia are identical with those of repairing any other hernia obliteration of the sac and reconstruction and replacement of the involved tissues

A nasogastric tube is inserted into the stomach before operation

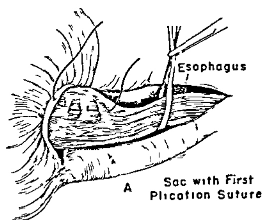
We prefer a thoracic approach through the bed of the left eighth rib or through the seventh or eighth intercostal space in younger patients The pulmonary ligament is divided and care is taken to clamp and ligate the vessel which is always found in it The mediastinal pleura overlying the esophagus is divided The latter is cleaned and defined and the vagus nerves are identified to avoid their accidental division

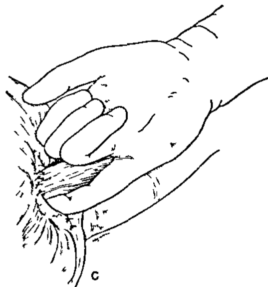
Hiatal hernia

A soft wet empty rubber wick is then passed about the esophagus for traction If a small sliding hernia is encountered it is best treated by simple plication of the sac in one or more layers as described by Sweet (37) (Fig -3) Several stitches of heavy silk are then used to approximate the limbs of the right crus of the diaphragm behind the esophagus Care should be taken not to tie these sutures too tightly The approximation of these fibers should be such that the little finger can easily be slipped between the crus and the esophagus (with a nasogastric tube inserted) Usually two or three sutures suffice

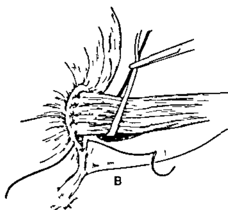
In the case of a larger sliding hernia an incision is made in the diaphragm after the esophagus is mobilized and a finger is passed up from below into the hiatus The hernial sac is opened and excised The rubber wick about the esophagus is passed down through the hiatus and gentle traction on this will reduce the esophagus well into the abdomen

The cut edges of the hernial sac are then sutured to the ab





The Little Finger Should Slip
Easily between Crura and Esophagus



SMALL SLIDING HERNIA

FIGURE 23 Repair of sliding hiatus hernia
(A) The hernial sac is reduced by plication If the sac is large more than one plication stitch is utilized and the last one only catches the muscular diaphragmatic ring (B) When reduction is complete the esophagus is tacked to the edges of the hiatus and the crura approximated (C) Care is taken not to approximate the crura too snugly A small gap should be left so that with the nasogastric tube in place the little finger can be passed through the hiatus

dominal peritoneum on the underside of the diaphragm the hiatus is repaired as previously described and a few fine stitches are used to anchor the esophagus to the edge of the hiatus above

In making this (or any other) diaphragmatic incision the operator should be cognizant of the usual distribution of the phrenic nerve in the diaphragm (5 26 31 34) The incision here extends

anteriorly, parapericardially, from the hiatus through the tendinous portion, to avoid a permanent segmental paralysis. A paracrural incision from the posterior attachment of the diaphragm just lateral to the fibers of the left crus directly forward to the central tendon is also safe and provides good exposure (31). Such considerations are not minutiae, but may be of considerable importance in older patients where most reliance is placed on diaphragmatic respiration with little costal action.

The mediastinal pleura is repaired with interrupted stitches of fine silk and the posterior gutter is drained for 24 to 48 hours by a large Foley catheter or a #30 colon tube on suction. In some cases drainage may be omitted.

We have avoided crushing the left phrenic nerve as a method of therapy. Too often this crush is permanent and not temporary. An upward displacement of the stomach may then occur sometimes resulting in torsion (5). These patients may have symptoms of the original hernia. Since the left phrenic nerve supplies the left half of the right crus (12), denervation of this portion of the hiatal musculature may increase cardiac incompetence and aggravate symptoms. Elevation of the diaphragm also results in undesirable loss of respiratory reserve. Drake (14) has found this procedure of value in persons whose general physical condition precludes a major operative procedure. Uebermuth (40) has advocated gastropexy in this group.

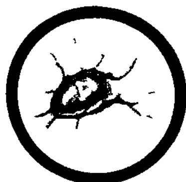
In the fragile elderly poor-risk patient who will not tolerate surgery, pneumoperitoneum may be a valuable emergency procedure. The elevation of the stomach and diaphragm and resultant narrowing of the esophageal hiatus may control bleeding and symptoms. Such a pneumoperitoneum is not deleterious to respiration in this elderly emphysematous group and may in fact reduce the amount of residual air and enhance ventilatory efficiency (24).

Most sliding hernias with shortening secondary to esophagitis can be reduced into the abdomen by the previous maneuvers. Freeing a greater length of esophagus proximally may aid in reduction. Occasionally one of the vagi is found to be taut, dividing





P1 Peritoneal sac



BENIGN TUMOR OF ESOPHAGUS



"JACK" IN ESOPHAGUS

PLATE VIII. Thoracic stomach, benign tumor of the esophagus, foreign bodies in the esophagus. (Courtesy Fletcher D. Woodward, M.D., Clinical Symposia, 8:3, 1956.)

diaphragm (Fig 24) A separate exploratory incision in the diaphragm can be made whenever necessary The hiatus will be found normal except in the unusual situation of combined hiatal and parahiatal hernia It is this type of hernia that occasionally *strangu*

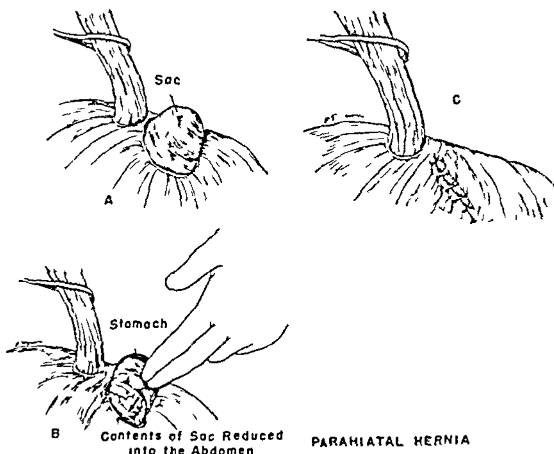


FIGURE 4 Repair of parahiatal hernia
(A) *Parahiatal hernia showing peritoneal sac* (B) Reduction of hernial contents after excision of sac (C) Repair of rent in diaphragm.

lates and requires emergency surgery There may occasionally be *an associated fibrous band obstructing the* stomach and duodenum in these patients and this should be looked for at the time of surgery

The sac is freed and excised and its contents are *reduced* into the abdomen The diaphragm is repaired with interrupted stitches of heavy silk Occasionally it may be advisable to use a strip of

fascia lata to reinforce the suture line or help close an unusually large defect

Repair of parahiatal hernia is usually technically easy and has the lowest recurrence rate of any esophageal hernia repairs

Hiatus hernia in children

During childhood and infancy boys are more often affected than girls Symptoms often begin during the first week of life and in almost all cases during the first year Vomiting is the commonest symptom and pyloric stenosis the chief condition in the differential diagnosis Bleeding occurs frequently

Conservative management coupled with a thorough understanding of child-parent relationships is frequently effective When symptoms persist beyond the first year and a hiatus hernia has been demonstrated by x ray, surgery is indicated Robb (33) has reported his experience with this group Of [illegible]9 infants and children with congenital hiatus hernia, 19 had simple hernial repair with 1 death 8 had esophagogastrostomy or esophagojejunostomy with 3 deaths 5 were treated conservatively and all survived Some patients required more than one operation

The hiatal hernias with short esophagus and thoracic stomach occasionally seen in children frequently become asymptomatic after the age of 3 or 4 (6) Bouginage is particularly effective in this group All 9 of the patients thought to have congenitally' short esophagus in Olsen and Harrington s series ([illegible]9) were treated by dilatation over a thread Franklin (18) reported the case of an 18 month old child with dysphagia and regurgitation whose x rays showed a short esophagus with stricture Treatment was bouginage Within [illegible] years roentgenograms showed the stomach lying completely in the abdomen and the esophagus appeared of normal length Husfeldt *et al* ([illegible]) are dissatisfied with conservative management In 11 patients so treated none was symptomless and all were still troubled by vomiting They recommend freeing the esophagus and stomach reducing the cardia below the dia-

phragm, and approximation of the crura In 10 cases of operation followed 2 to 15 months, 7 patients were symptom-free 1 had a recurrence, and in 2 operation was not technically feasible Mobili-

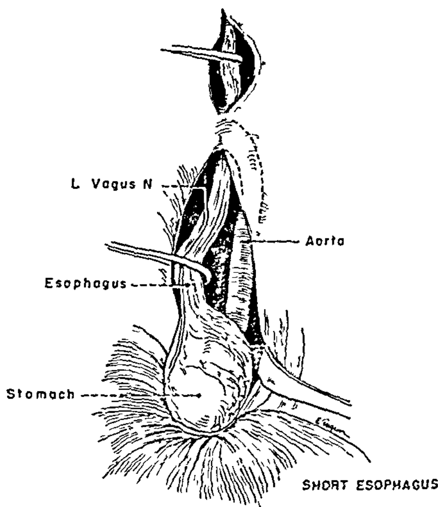


Figure 5 Extent of mobilization which may be necessary to achieve reduction of the cardia below the diaphragm in case of short esophagus *Particular care is taken not to injure the vagi*

zation of the entire esophagus above the arch of the aorta to the apex of the chest may be necessary to permit such reduction (Fig 5) *The phrenic nerve is never crushed and* particular care should be taken to provide and maintain lung expansion

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Esophageal Ulcer

10

Peptic ulcer of the esophagus is an ulcer of the mucous membrane of the esophagus which is caused by the action of acid and pepsin contained in gastric juice.

Esophagitis is always present in cases of esophageal ulcer. Many of our cases have been associated with hiatus hernia, duodenal ulcer, benign stenosis of the esophagus, and congenitally short esophagus. In a number of cases a tube of gastric epithelium lines the lower end of the esophagus. It seems probable that esophagitis with erosions often precedes the development of a true peptic ulcer.

Incidence

The incidence of esophageal ulcer is very much less than that of peptic ulceration in the stomach or duodenum. Jackson (4) reports the finding of peptic ulcer of the esophagus at [illegible] per cent of all esophagoscopies. MacMillan (5) states that esophageal ulcer accounts for 1.5 per cent of all pharyngeal and esophageal disease. Peptic ulcer of the stomach and duodenum is probably about [illegible]0 times as common as peptic ulcer of the esophagus. It should be made clear that we are omitting erosions and superficial ulcerations

because we do not believe that they represent true peptic ulcer of the esophagus We believe that the variability in definition accounts for the discrepancies among reports Peptic ulcer of the esophagus usually involves the mucosa somewhere in the lower 10 cm It may occur however in the middle or upper esophagus It is at least 5 times more common in men than in women and is more likely to occur in middle life

Etiology

The most likely cause is regurgitation of acid peptic secretions into the lower portion of the esophagus The fact that most esophageal ulcers occur in that area tends to substantiate this theory as does the frequent concomitance of hiatus hernia There must however, be many persons in whom regurgitation occurs who do not develop significant esophagitis or esophageal ulcer

The finding of a tube of gastric mucosa lining the lower esophagus would indicate that heterotopic gastric mucosa might be an important etiologic factor, however if the mucosa is gastric the ulcers must also be gastric and not true esophageal ulcers Ulcers in the mid and upper esophagus may develop in islands of heterotopic gastric mucosa Peptic ulcer of the esophagus also may occur at a surgical anastomosis between the esophagus and the stomach, in fact esophageal ulcer may be produced by any state which will allow acid gastric secretions to come into prolonged contact with the esophageal mucosa

An attempt has been made to explain a high ulcer in the esophagus on the basis of the marked kinking of the esophagus which might occur in kyphoscoliosis (3) however in a case of kyphosis with which we are familiar there was not much kinking but there was an island of ectopic gastric mucosa

The fact that dietary deficiency might play a part has been indicated by Brown (4) During a lengthy course of feeding a deficient rice diet to white rats Brown frequently noted lesions of the esophagus consisting of focal ulcerations of the mucosa with

penetrating inflammation When synthetic vitamins were added to the diet, well in excess of the vitamin requirements the rats gained in weight, nearly all the ulcers healed, and at autopsy the animals showed practically no lesions in the internal viscera

Pathology

Ulceration of the esophagus is a circumscribed lesion with loss of mucosa and inflammatory reaction It usually takes the form of a single ulcer If the ulceration is close to a blood vessel hemorrhage may occur If it penetrates deeply, perforation may take place Long-continued ulceration often progresses to chronic fibrosis with moderately severe stenosis When marked fibrosis and severe stenosis have developed the situation cannot be reversed by medical treatment and bouginage, hence surgery is necessary On the other hand in relatively superficial esophageal ulcers there may be only a moderate amount of surrounding inflammation and the lesion may heal almost completely without fibrosis and stenosis These are the cases that respond well to treatment by diet and bouginage

Symptoms

The usual symptoms of esophageal ulcer are pain, dysphagia odynphagia and vomiting

Pain The pain of peptic ulcer of the esophagus is usually substernal subxiphoid or high epigastric, and is often accompanied by heartburn and distress It may occur at any time of day or night and it is much less consistently related to the time of food intake than is the pain of gastric or duodenal ulcer It may be considerably relieved by milk soft bland foods and antacids but complete relief through such a regimen is less likely in this condition than in gastric or duodenal ulcer

Dysphagia (difficulty in swallowing) Most patients with esoph

ageal ulcer have some esophageal obstruction and retention of food in the esophagus If one asks such a patient whether or not he has difficulty in swallowing he may reply in the negative and unless one asks further questions he may not reveal the fact that solid food remains in the esophagus As time goes on the inflammatory process may become more acute or fibrous tissue may form to such a degree that the patient also has difficulty in swallowing liquids

Odynphagia (painful deglutition) This is a rather common symptom of esophageal ulcer The pain is apparently associated with the passage of food over the ulcer Coarse or spicy foods are more likely to cause pain than is a soft bland diet

Vomiting This is a common symptom of esophageal ulcer and is probably due to the irritating effect of the ulcer and the esophagitis in combination with the stenosis almost invariably produced by the healing process It may be more regurgitation than actual vomiting When there is incompetence of the cardiac sphincter regurgitation occurs especially during recumbency stooping or straining Any situation which increases intra abdominal pressure such as obesity or the wearing of tight clothing around the abdomen aggravates the symptoms

Hemorrhage from peptic ulcer of the esophagus may occur in small amounts in the form of melena but is usually not a prominent symptom We know of two cases in which spontaneous hemorrhage was so severe on several occasions that surgical resection was indicated The operation was successfully performed in one case during a quiescent period but in the other case that of an elderly poor-risk patient with active pulmonary tuberculosis and an actively bleeding esophageal ulcer resection was carried out as an emergency procedure with a fatal result (In the latter case however the surgeon s hand was forced) We also know of two cases of moderately severe bleeding following esophagoscopy and bouginage

Loss of weight is usually noted in this condition it seems to be entirely due to the patient s inability to eat an adequate diet

Complications

Esophageal ulcer may be complicated by hemorrhage, perforation, or obstruction Minor episodes of bleeding probably occur with some frequency but may go unrecognized Moderate or severe hemorrhage is rare but is likely to be an indication for surgery Perforation also is rare, but a fatal case of perforation of the aorta by a benign esophageal ulcer was reported by one of us (G L N) in 1953 (6) The healing of an esophageal ulcer frequently leads to cicatricial stenosis and varying degrees of esophageal obstruction



Figure 6 Small constant niche (showing in x ray just above small hiatus hernia) about 4 mm in diameter which was described by the radiologist as probable esophageal ulcer Esophagoscopy however disclosed only severe esophagitis with multiple hemorrhagic erosions

Diagnosis

X-ray examination

The radiologist is likely to suspect peptic ulcer of the esophagus if he observes esophageal spasm, delayed passage of barium, narrowing of the lower esophagus, hiatus hernia, or short esophagus. The demonstration of an ulcer crater, however, is difficult, and requires the taking of many spot films to show constant pooling of barium in one location (Figs. 26, 27). Although such constant pooling is characteristic of an ulcer crater (Fig. 28), it sometimes occurs between the esophageal folds and this finding may prompt an erroneous diagnosis of ulcer. There are also cases in which the



Figure 27. Roentgenogram showing pooling of barium at lower end of the esophagus with stricture. X-ray diagnosis: benign stricture, hiatus hernia, esophageal ulcer and duodenal ulcer. History of heartburn since childhood; 3 years regurgitation and substernal pain uncontrolled by diet, as well as by esophagoscopy and bougienage. As is frequently the case, the stricture was above the ulcer, preventing endoscopic view of the ulcer. Severe hemorrhage later made resection of the lower esophagus necessary.

radiologist has suspected benign peptic ulcer but the final diagnosis was either achalasia (Fig 29), foreign body (Fig 30), or carcinoma (Fig 31)



FIGURE 28 X ray appearance of esophageal ulcer demonstrating pool of barium in the ulcer crater Above the ulcer there is a benign stricture This was confirmed by esophagoscopy and although the ulcer was not visualized biopsy revealed peptic ulceration

Sometimes the esophagus appears to be short, and the esophagogastric junction may be located at various levels above the diaphragm — even as high as the aortic arch Gastric folds may be seen extending through the diaphragmatic hiatus into the thorax When the patient is examined in the Trendelenburg position reflux from the stomach may be demonstrated

No x-ray study of the esophagus is complete without careful rotation of the patient in all directions and the taking of numerous spot films The outlining of esophageal folds and gastric folds in a hiatus hernia is important If an esophageal ulcer niche is large, it *may resemble a diverticulum*



FIGURE 29 Roentgenogram showing ulcer crater with constricting lesion of the esophagus above hiatus hernia Esophagoscopy showed the narrowing but no ulcer was visible Biopsy however from deep within the lumen of the stricture caught the edge of the ulcer Pathology report peptic ulcer of the esophagus normal gastric mucosa.

Esophagoscopy

Esophagitis without true peptic ulceration is a fairly common disease which may give rise to the same symptoms as peptic ulcer Esophagoscopy is indicated for differential diagnosis when the



FIGURE 30 Pooling of barium suggestive of peptic ulcer of the esophagus but later proven by esophagoscopy to result from a portion of a dental plate stuck at the lower end of the esophagus—successfully removed by esophagoscopy

FIGURE 31 X-ray of patient who for 19 years was treated for benign stricture and peptic ulcer of the esophagus Diagnosis at this hospital through esophagoscopy with biopsy demonstrated carcinoma The importance of biopsy cannot be overestimated

multiple erosions and superficial ulcerations of the esophagus which occur in association with esophagitis are present A true peptic ulcer may be difficult to demonstrate by esophagoscopy because frequently there is considerable stenosis above it which prevents an adequate view of the lesion Allison (1), however, found x-ray evidence of esophageal ulcer in only 5 of 74 cases in which an ulcer was demonstrated by esophagoscopy, but our experience has convinced us that radiology is of great importance in the diagnosis of peptic ulcer of the esophagus If there is no stenosis and no suspicion of carcinoma, the clinician may not even ask an endoscopist to examine a case of uncomplicated peptic ulcer of the esophagus In such cases the condition may be suggested clinically and roentgenologically and may then be treated medically without

further diagnostic study We believe however that all such cases should be examined by esophagoscopy and an adequate biopsy specimen taken

In our experience the endoscopist is usually called upon to examine and treat stenosis of the esophagus caused by a peptic ulcer located just below the narrowing In such a case the ulcer may not be visible by esophagoscopy On the other hand in some cases a biopsy specimen taken from deep within the lumen of the stricture may catch the edge of an ulcer with the result that the pathologic report offers conclusive proof of the presence of a peptic ulcer of the esophagus In any case regardless of the suspicion of carcinoma esophagoscopy should always be performed in order to obtain a biopsy specimen and to ensure that an unsuspected carcinoma is not missed Saline washings are usually taken from the suspected area for cytologic examination A smooth cone like narrowing usually indicates a benign stricture but there are some smooth annular carcinomas which present much the same picture In such cases a biopsy specimen taken from deep within the narrowed area is essential to a positive diagnosis A positive cytologic report under such conditions also would be of considerable importance but a single positive cytologic finding should not be regarded as conclusive if it is the only indication of carcinoma

In some cases of achalasia ulceration of the esophagus has been reported by the radiologist In such instances esophagoscopy must be performed to differentiate peptic ulcer and achalasia In the latter condition esophagoscopy usually reveals a dilated esophagus with no ulceration and no narrowing In peptic ulcer of the esophagus however the procedure often demonstrates marked inflammation and considerable stenosis

TREATMENT

Medical treatment

Diet A soft bland diet consisting of strained or finely ground food given in small amounts at frequent intervals is the most im-

portant factor in successful medical management. Alcohol and tobacco should be completely eliminated. Strongly acid or highly seasoned foods are prohibited. In some cases there is so much inflammation or narrowing of the esophageal lumen that the patient can tolerate nothing but liquids. At the onset, frequent milk and cream feedings should be given.

For cases in which inflammation or narrowing is severe the following schedule is recommended:

7 A.M.	6 oz.	sweetened orange juice
8	10 oz.	cereal water*
9	6 oz.	chicken broth †
10	6 oz.	malted milk
12 NOON	8 oz.	cream of pea soup
1 P.M.	6 oz.	cereal water
2	6 oz.	banana milk shake
3	6 oz.	bouillon plus beef juice †
4	6 oz.	eggnog
5	8 oz.	oyster stew (strained) †
6	6 oz.	orange juice
7	8 oz.	cream of tomato soup
8	6 oz.	cocoa

This diet contains approximately 3000 calories (carbohydrates 350 gm., proteins 75 gm., fats 150 gm.).

When the patient can tolerate a little more food, other items may be added, all foods being strained through a #25 sieve. A suggested meal plan on this diet would be as follows:

7 A.M.	Strained cereal with milk or cream, cocoa
8	Strained orange juice with sugar
10	Cereal water
12 NOON	Creamed minced chicken
	Strained spinach with butter and cream

* Cereal water is made with 1 cup of water, 1 tablespoonful of flour (barley, rice or wheat) and tablespoonfuls of sugar.

† Chicken broth, bouillon plus beef juice, and oyster stew (strained) may be excluded in the early stages of esophageal ulcer management since they act as secretagogues; milk and cream, soft boiled eggs, strained cereal or liquid gelatin may be substituted.

	Milk with part cream
	Soft custard pudding
2 P M	Chocolate malted milk
4	Tomato juice 6 oz plus beef juice or liver pulp oz.
5	Welsh rabbit (no bread or crackers)
	Strained beans milk with cream strained peaches
8	Eggnog with part cream

This diet contains approximately 3100 calories (carbohydrates, 350 gm proteins 85 gm fats 150 gm)

Medication Relief of pain may be obtained in some cases through the use of sodium bicarbonate aluminum hydroxide bismuth subnitrate calcium carbonate bismuth and magnesium oxide and other antacids Belladonna is rarely helpful in the management of uncomplicated esophageal ulcer but may be useful in the treatment of an accompanying duodenal ulcer with pylorospasm

Patients should be advised to sleep with the head of the bed elevated about six inches so that the force of gravity may be utilized to keep acid gastric secretions in the stomach Obese patients must reduce weight must not wear tight clothing and should avoid stooping

Endoscopic treatment

Bouginage through the esophagoscope is of very great importance in the management of the narrow lumen which frequently accompanies esophageal ulcer It should be performed only by an experienced esophagoscopist In some cases bouginage may be carried out by means of a special bougie with a previously swallowed thread being used as a guide (see Chapter 6 Methods of Treatment)

Endoscopic packing of the esophagus has been suggested as a method of stopping hemorrhage from esophageal ulcer We do not use this technic since we believe that it is too temporary and prefer more definitive methods Furthermore we avoid endoscopic cauterization of bleeding areas — not only because the method is

Benign Peptic Stenosis

II

Benign stenosis of the esophagus is a general term which denotes narrowing of the esophagus. In this chapter we deal entirely with so called peptic stenosis due to regurgitation of acid peptic secretions from the stomach. We are postponing the discussion of web like stricture and achalasia to later chapters, since they are separate diseases unrelated to the stenosis of peptic esophagitis or ulceration.

Incidence

Increasing interest in esophageal disease and improved methods of radiologic and endoscopic diagnosis have resulted in greater awareness of this condition.

In a series of 1000 esophagoscopies (7) 60 cases of benign stenosis were encountered, in 18 of which there was esophagitis alone, in 34 hiatus hernia, in 20 duodenal ulcer, and in 16 esophageal ulcer. Six of these patients had all three conditions: hiatus hernia, duodenal ulcer and esophageal ulcer. None had ulcer in the abdominal portion of the stomach. Forty two were male and 18 female. Two patients were only 15 years old, 1 was 25, 17 were between 30 and 50, and 36, or 59 per cent, were between 50 and 70. Four patients were between 70 and 80 and 1 was 82. Further studies

of additional patients have been reported by Benedict and Gillespie (4, 5) In a study of 1340 patients examined in this clinic they found 119 (8.7 per cent) cases of peptic stenosis Lodge (11) found microscopic evidence of acute esophagitis in 31 per cent of 500 unselected hospital patients and in 3 per cent of 100 patients who died quickly in accidents

Etiology

All patients with benign stenosis have associated esophagitis We believe that the esophagitis precedes the development of the stenosis and is an important factor in its etiology (6) Regurgitation of acid peptic secretions undoubtedly plays the most important role in the development of esophagitis and esophageal ulcer Hiatus hernia is a very common predisposing cause since it makes regurgitation more likely In our series 85 per cent of the patients had a hiatus hernia Forty per cent showed evidence of esophageal ulcer on routine examination 40 per cent also showed x ray evidence of present or past duodenal ulcer One hundred and nineteen biopsy specimens were taken from 80 patients 49 of these demonstrated gastric mucosa at one or more biopsies (6[illegible] per cent) and the remainder showed acute and chronic inflammation of the mucosa If a duodenal ulcer is present it may mean hyperacidity and pyloric obstruction with increased vomiting and regurgitation Six patients with acute esophagitis were heavy users of alcohol and 6 used alcohol moderately Fourteen patients had suffered from gallbladder disease at some stage of their lives and many of these had had a great deal of vomiting which may have contributed to their esophagitis and stenosis Twenty-two patients had had a surgical operation requiring anesthesia and hospitalization within 5 months of the development of dysphagia In 5 cases an indwelling gastric tube was used but this is probably not statistically significant as Levin tubes are commonly used without causing the development of enough esophagitis to cause stenosis

As early as 1906 Tileston (17) held that insufficiency of the

cardia which allows regurgitation of gastric juice, was essential for the development of the esophageal ulcer and hence that vomiting — as in peritonitis, duodenal ulcer, and pyloric stenosis — was important in its etiology

Winkelstein (18) in 1935 described 5 cases of a new clinical entity which he called "peptic esophagitis" and stated that the course of the disease the location of the ulcers the concomitance of peptic ulcers the hyperchlorhydria, and the fact that relief could be obtained through ulcer treatment suggested a peptic origin of the esophagitis

Allison (1) in a significant paper on 74 cases of peptic ulcer of the esophagus considered that a hiatus hernia which allowed regurgitation of acid gastric juice was the essential factor Olsen (13) regarded reflux of gastric juice as the most common cause of esophagitis and stressed the effect of vomiting and incompetence of the cardia from such conditions as hiatus hernia

In 1936 Selye (15) described the production in rats of what he called peptic hemorrhagic esophagitis by means of pyloric ligation which he believed to indicate that the presence of gastric juice for an abnormal time in a normal esophagus would produce inflammation

Ferguson *et al* (9) in experiments on various types of animals found that acid gastric juice which remained in contact with the esophagus for an abnormal time whether owing to vomiting or regurgitation or as a result of direct application had a devastating effect whereas hydrochloric acid in a concentration similar to that in gastric juice had little effect by itself These workers believe that non specific esophagitis is due primarily to the action of acid peptic juice'

PATHOLOGY

Peters (14) has published an excellent and very thorough review of the pathology of severe digestion esophagitis She discusses the etiology incidence and morbid anatomy of 116 severe cases Ac

cording to Peters in the acute stage the mucosa is invariably shed to some extent discrete shallow ulcers with brown or blood stained bases may be seen occasionally round or oval they are more usually linear and follow either the summits or the folds of the lower closure pleats

In more severe cases the whole or part of the tube is mostly hemorrhagic often the whole tube is stripped of epithelium except for scraps at the two ends

Peters describes the chronic type as follows As in the previous group the appearances are very variable any length of the tube may be involved but in the older cases the changes are more likely to be concentrated at the lower end In the more recent (active chronic) there is dirty brown rough ulceration in patches and streaks or involving the whole circumference characteristically with islets of surviving epithelium in or above the ulcerated area Extensive regeneration occurs from the margins of these sometimes appreciable to the naked eye as a delicate hazy film covering adjacent raw surfaces The islets vary in shape size and colour, the more recent are pinhead to lentil sized rounded and grey-white bigger and older patches are flattened scaly white plaques like snakeskin or thick strips like candle grease gutterings separated by irregular channels of superficial ulceration These changes may persist in or above the most chronic lesions or become part of an extensive leukoplakia which occasionally progresses to cancer Active fibrosis begins at this stage and advances with time

In the second probably more frequent form the appearances are conditioned by great fibrosis As the scar tissue contracts two important secondary changes develop Concentric contraction gives permanent stricture and in the case records the gradual conversion of intermittent spasm dysphagia to constant obstruction may be clear Longitudinal contraction leads to permanent slight shortening which following spasm shortening makes the previously sliding hernia irreducible and ensures permanent incompetence of the pinchcock It is probably the commonest cause of permanent shortening of the gullet

Symptoms

Dysphagia is the commonest and most important symptom of peptic stenosis of the esophagus It may vary from slight difficulty in swallowing solid food to complete inability to swallow liquids Regurgitation is common and usually occurs without nausea True vomiting is uncommon Heartburn is a frequent symptom as is substernal or subxiphoid pain or distress especially if there is ulceration Anorexia may be present Patients frequently note an inability to belch When regurgitation occurs the material may be bloody, especially if there is severe inflammation or ulceration

Complications

Benign peptic stenosis of the esophagus may be complicated by hemorrhage perforation or carcinoma As a rule hemorrhage is only slight, and is due to the associated esophagitis, however if more and more ulceration occurs, moderate or severe hemorrhage may take place

Perforation is uncommon in benign peptic stenosis but it may occur if the accompanying esophagitis progresses to deep ulceration

As regards carcinoma we know of one man who was treated over a period of 19 years for benign peptic esophagitis with stenosis but was later proved to have carcinoma in the same area That patient had received an early diagnosis of hiatus hernia esophageal ulcer and duodenal ulcer and he had been treated for those conditions in leading clinics of the United States and Europe (See case report M D page 74) Incidentally his case was reported by Levine (10) in 19-9 as a case of peptic ulcer We believe this to be very rare indeed

Diagnosis

The diagnosis of benign peptic esophageal stenosis may be suspected on the basis of a history of dysphagia substernal pain and

regurgitation The history may show that symptomatic relief has been obtained through a bland liquid or semi-solid diet or through the use of antacids X-ray examination is of fundamental importance it usually reveals a smooth funnel-shaped narrowing of varying length through which the barium passes slowly if there is much obstruction Sometimes there is complete obstruction — in which case no barium passes If obstruction is slight there may be no delay in the passage of barium Sometimes the radiologist is unable to differentiate benign stenosis and carcinoma In all cases esophagoscopy is indicated

Esophagoscopy

Direct inspection of a benign inflammatory esophageal stenosis usually *reveals a smooth conical narrowing with varying degrees* of inflammation depending on the stage of the disease (Plate VI ±6) Erosions and superficial ulcerations are frequently seen especially if the disease is very acute Increased reddening is nearly always present and there is usually some edema Denudation of the mucosa is often noted The inflammatory process may be slight and be localized close to the stricture or it may be marked and may extend 10 or 15 cm above the narrowed area Sometimes a vegetable foreign body or particle of meat is found lodged in the stenotic zone and this contributes markedly to the esophageal narrowing Any such foreign body must be removed bouginage carried out and a biopsy specimen obtained in order to exclude carcinoma The site of the biopsy is of very great importance The specimen should usually be taken from deep within the lumen of the stenotic sector in order to obtain a representative specimen However it should be borne in mind that the forceps may be inserted even beyond the stenosed area and hence a non cancerous specimen obtained when the neoplastic tissue is just proximal to the end of the forceps This however happens rarely — and only when the stenotic sector is very short The biopsy must be taken with great care to avoid perforation If a pathologic report of chronic inflamma

tion is received and carcinoma is still suspected, esophagoscopy and biopsy should be repeated

Cytology

Cytologic study of esophageal washings is important but in our experience this procedure has only once revealed a carcinoma of the esophagus which was not suspected after x ray or esophagoscopy This occurred in a case of achalasia in which x ray examination and esophagoscopy indicated achalasia and were negative for carcinoma (See Chapter 4 case reports) In this case the carcinoma was only 1 cm in diameter and was difficult to find at the time of surgery In a series of 70 patients studied by this method at this hospital there have been two false-positive examinations

TREATMENT

Dietary treatment

The esophagus is continually subjected to trauma by the passage of food and drink Unwise eating or drinking may be highly injurious to normal esophageal mucosa and is certainly very traumatic to an already inflamed mucous membrane Sometimes patients with severe inflammatory stenosis of the esophagus should at first receive intravenous alimentation and be given nothing by mouth Later on bland liquids and thin cereals may be tolerated Some fruit juices may be irritating When the narrowing is not too marked the patient may tolerate strained vegetables and ground meat Alcohol and fried or highly seasoned foods should be entirely omitted

Medicinal therapy

Antacids are useful in the treatment of peptic esophagitis with or without stenosis Aluminum hydroxide is excellent in either liquid or tablet form such as Amphojel Creamalin or Gelusil Sodium

bicarbonate also is useful but cannot be used over a long period since kidney stones may form Occasionally there is some spasm in inflammatory diseases of the esophagus this may be ameliorated by the use of nitroglycerin, grains 1/200 placed dry under the tongue just before a meal Atropine sulfate usually has little effect in esophageal spasm

Bouginage

Except in certain intractable cases the treatment for benign stenosis of the esophagus is bouginage Esophagoscopy should always be carried out in every such case both in order to observe the degree of inflammation and stenosis and in order to carry out bouginage under direct vision and to obtain a satisfactory biopsy specimen When carcinoma has been excluded and the diagnosis of benign peptic stenosis established further bouginage may be carried out in the office or in the out patient department with a previously swallowed thread being used as a guide (see Chapter 6)

Surgical treatment

The indications for surgery are obstruction intractable pain hemorrhage and perforation — *i e*, failure of conservative management This latter is often difficult to define The older poor risk patient may subsist indefinitely on a regimen of soft bland food and periodic dilatations — a program not usually acceptable to a younger or more active person

Plastic procedures

Rarely one may encounter a sharply localized stricture or area of stenosis which does not respond to bouginage but is benefited by conservative local operations Esophagoplasty consisting of vertical incision and transverse closure (Fig 3-A) has been advocated as a method of dealing with these localized conditions If the nar-

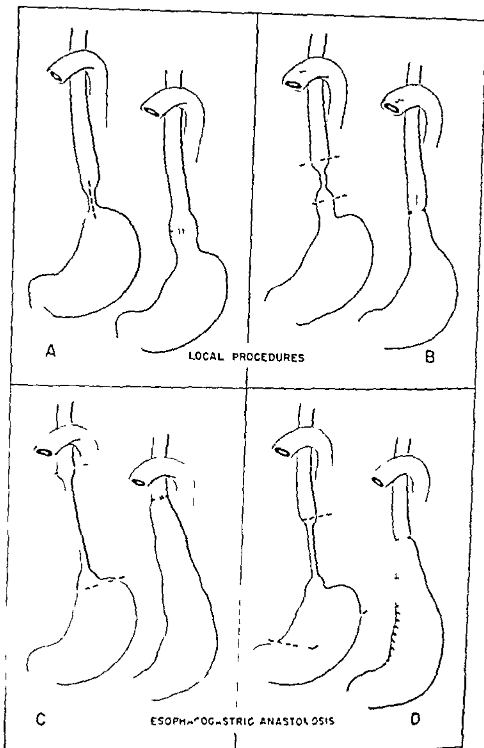


FIGURE 32 Surgical procedures in benign peptic stenosis
(A) Esophagoplasty with vertical incision and transverse closure (B) Local resection with end to end anastomosis (C) Subaortic or supra aortic anastomosis (D) Subaortic anastomosis with resection of large stomach segment

rowed area is so located that this maneuver can be performed without incision of the cardia and loss of esophagogastric competence, very satisfactory results can be obtained On the other hand an esophagoplasty which is extended through the cardia into the stomach may frequently cause distressing complications which are worse than the original disease (see discussion of cardioplasty in Chapter 14)

Some well-localized strictures can be resected locally with end-to-end anastomosis (Fig 5-B) In this maneuver it should be remembered that several centimeters of normal esophagus must be mobilized at either end and that meticulous care and judgment must be exercised to maintain an adequate blood supply to the anastomosis

Resective procedures

We have found that most esophageal strictures for which surgery is indicated require resection with esophagogastric anastomosis Usually such a resection must be extensive if uninvolved esophageal tissue is to be obtained for the anastomosis An extensive resection also may be of prophylactic value since in the high anastomoses postoperative esophagitis is relatively infrequent It is possible that the greater number of mucus glands present in the upper esophagus provide more protection against reflux

A left transthoracic approach through the bed of the eighth rib is utilized If further exposure upwards should be needed the seventh or sixth ribs may be divided posteriorly — or preferably the fourth rib may be resected this method produces a second operative incision which will greatly facilitate and increase the safety of a high anastomosis

The mediastinal pleura is incised and the esophagus mobilized The diaphragm is divided through the esophageal hiatus and the stomach mobilized (see section on hiatus hernia for hazards in sectioning diaphragm and chapter on carcinoma of esophagus for operative details and illustrations) The esophageal dissection may be

difficult because of vascular adhesions secondary *to local inflammation*. The esophagus should be transected well beyond any grossly visible abnormal mucosa. Failure to remove all inflamed tissue will result in recurrence of the stenosis. Occasionally a *supra-aortic* anastomosis may be required (Fig. 3-C).

When the extent of resection is such that a subaortic anastomosis can be accomplished, an alternate and possibly better procedure is resection of a much larger segment of the stomach to obtain maximal reduction of gastric acidity (16) (Fig. 3-D).

Patients with long established esophageal stricture and irreversible fibrosis frequently have a coexisting duodenal ulcer with pyloric obstruction (3). The vagotomy which is an unavoidable factor in esophageal resection causes gastric retention which when associated with pyloric obstruction is particularly distressing. The concomitant execution of simple pyloroplasty may eliminate these complications.

Other procedures

The variety of operative procedures reported in the literature attests to the lack of a uniformly successful surgical treatment of this condition. In the interest of reasonably thorough coverage of the subject, some of these procedures — *with which we have had* no personal experience — will be briefly mentioned.

McLean and Wangensteen (12) have treated 15 patients with esophageal resection by distal subtotal gastrectomy and 3 by combined hiatus hernia repair and pyloroplasty. Results were excellent in all cases.

Excellent results also have been obtained by esophagojejunostomy performed in a Roux-Y fashion, with preservation of most of the stomach (2, 8).

Prognosis

The prognosis in benign stenosis of the esophagus is very *good* when palliation by bouginage is attempted. Complete cure by this

method is probably infrequent since some narrowing nearly always remains, as can be demonstrated by x-ray examination However many patients are able to eat most foods after a series of such treatments Surgery, when carried out with meticulous care particularly with respect to the anastomosis usually gives good results but in some cases inflammatory stenosis has recurred at the esophagogastric anastomosis In such cases further bouginage may be necessary

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Caustic Stenosis

12

The term caustic stenosis of the esophagus means a burn caused by a corrosive agent with enough scar formation to cause some esophageal obstruction

Etiology

Caustic stenosis is caused by the swallowing of strong alkali or acid solutions In 95 per cent of cases the condition is due to the ingestion of commercial lye solutions (4) which are about 95 per cent sodium hydroxide (10) Household ammonia potassium carbonate (salts of tartar) sodium carbonate (washing soda) and bichloride of mercury also may produce esophageal stenosis Strong acids are occasionally responsible

Chronic acetanilid poisoning has caused marked annular narrowing of the esophagus as reported by Wolfson and Greenberg (25) Liquor adulterated with lye has been reported as a cause of dysphagia and stricture of the esophagus (1.) (Caustic soda is used in Finland for adulterating liquor)

Despite the fact that our laws require proper labeling of commercial cleaning substances containing caustics such products are often carelessly left about within reach of children It is quite

obvious too that many adults contemplating suicide believe that lye is a poison which will kill them, rather than one which will cause severe burns to the buccal, esophageal and gastric mucosa with resulting stricture formation

Pathology

The most severe burns usually occur in the middle and lower thirds of the esophagus but burns in the upper esophagus also are common According to Fatti Marchand and Crawshaw (7) there are four grades of stenosis 'according to their length and density *In Grade 1 fibrosis is circumscribed being confined to less than the* total circumference of the esophagus and to a short segment in length There are shelf strictures and they never cause significant narrowing of the lumen Grade 2 *these are localized annular strictures* involving the total circumference of the esophagus The fibrosis does not penetrate beyond the submucosa and consequently narrowing is never pronounced and treatment never a difficult problem Grade 3 these are dense strictures of limited length (arbitrarily less than ½ inch long) Fibrosis extends through the outer muscular coats and perhaps the lumen which is lined by epithelium may be reduced to a thread

Grade 4A These are strictures of more than half an inch in length which have little tendency to progressive narrowing The subgrade is judged by the fact that dilatation is easy and the lumen maintains its stretched caliber Fibrosis is confined essentially to the mucosa and submucosa The stricture is lined by epithelium regeneration having occurred from surviving islands of mucosa

Grade 4B these tubular strictures have densely fibrotic walls periesophageal adhesions and marked luminal narrowing Because of the extensive destruction epithelium cannot rebridge the gap and the lumen is lined by fibrous and granulation tissue Such strictures are always found to be difficult to treat

In these authors experience approximately 50 per cent (of 147 cases) of all patients admitted with acute caustic soda poisoning

develop esophageal strictures This does not imply that corrosive burns of the esophagus may heal completely but rather that in half the cases the corrosive never enters the esophagus

Sometimes a burn is deep enough to penetrate all the layers of the esophagus and cause a perforation with mediastinitis Before the advent of chemotherapy such burns with perforation were universally fatal

Symptoms

At the time of the accident the diagnosis may be obvious in view of the presence of burns on the lips mouth and tongue The patient or the family may give a history of possible lye ingestion Excessive salivation is likely For several weeks the patient may have no difficulty in swallowing but after the cicatrix begins to develop in the esophagus gradually increasing difficulty in swallowing solid food will be noted

Over 75 per cent of the patients are children less than 6 years of age Stenosis usually occurs within 2 months of the accident but sometimes is not evident until more than a year later

Complications

In addition to the complications which may result from treatment (page 128) — hemorrhage and perforation — one must consider *foreign body total atresia and carcinoma*

Foreign body In lye stricture as in other strictures esophagoscopy with direct inspection of the lumen may reveal a foreign body such as a bolus of meat which is actually the chief cause of the obstruction Removal of this foreign body is usually readily accomplished by inserting a suitable forceps through the esophagoscope

Total atresia of the esophagus may follow a severe lye burn (see page 129 for treatment)

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FIGURE 33 Complete obstruction from caustic stenosis of the esophagus.

Carcinoma Another complication of lye stricture of the esophagus is the possibility that carcinoma will develop (1, -) Up to 1941, 33 cases of carcinoma of the esophagus had been reported as developing in benign stricture, although Marchand (14) found none in 133 patients with caustic stricture Most of the carcinomas were noted after the ingestion of lye but in rare instances they were said to have been observed in cases of foreign body, cardiospasm, hysterical dysphagia, and congenital stricture It is probable that chronic esophageal irritation plays a part in the development of cancer in areas of stricture Carver *et al* (4) encountered only 2 patients with esophageal carcinoma in -33 cases of lye stricture

Diagnosis

Immediately after the accident the diagnosis may be made on the basis of history and physical examination as outlined above In the later stages the history is also important and x-ray examination reveals the level and degree of stenosis (Figs 33 34) If the stenosis is not complete x-ray examination may also demonstrate multiple strictures at different levels in the esophagus and in addition may show involvement of the stomach

Treatment

Esophagoscopy is of limited value in the diagnosis and treatment of caustic burns of the esophagus In the acute stage it is contraindicated because instrumentation may increase the damage already done and in the chronic phase the esophagoscopist can see only the upper margin of the most proximal stricture In a few cases esophagoscopy (Plate IX #1) and bouginage under direct vision is useful

Van den Ostende (-3) claims that esophageal strictures that result from the swallowing of corrosive substances can largely be prevented by treatment with cortisone This author reports only one case and we ourselves have not yet had an opportunity to use this



FIGURE 34 X ray appearance of caustic stenosis after bouginage Clinical relief after repeated bouginage was very good in fact much better than the x ray indicates

treatment Weisskopf (24) experimentally produced lye burns of the esophagus in dogs he found that strictures did not develop in the animals treated with cortisone, whereas they did appear in all animals in the control group

Acute phase

Immediate treatment consists in an attempt to neutralize the chemical. In the case of *alkalis*, vinegar should be given. If *acids* have been swallowed, sodium bicarbonate should be administered. In cases of acid or alkali ingestion the stomach tube should not be used. However, if *bichloride of mercury* has been taken, emptying of the stomach by inducing vomiting or passing a tube as soon as possible after the ingestion of the poison is the most important determining factor in recovery. This should be followed by gastric lavage with a 5 per cent solution of sodium formaldehyde sulfoxylate, of which 200 cc. should be left in the stomach. If this solution is not available, introduce one pint of milk and several eggs into the stomach; this procedure should be followed by lavage with a saturated aqueous solution of sodium bicarbonate. In all cases general supportive treatment is indicated, including external application of heat, morphine if necessary for pain, sedatives as indicated, ephedrine, 25 mm. administered subcutaneously, and Coramine, 5 cc. given intravenously as necessary for medullary stimulation, parenterally given fluids and blood transfusion for shock. Nothing should be administered orally as long as nausea and vomiting are present; later olive oil up to 8 oz. should be given by mouth. Bismuth subcarbonate taken orally is a good antidote for burns in the mouth or the esophagus. As soon as it can be tolerated, water should be given by mouth.

Chronic phase

Very soon after the accident a swallow of thin barium should be given to determine the amount of immediate esophageal damage. If it appears that very little damage has occurred, one must beware of the latent period which sometimes extends over several weeks or even months before fibrosis and strictures occur. During this period it is advisable to have the patient swallow a thread as soon as possible

in order to maintain the patency of the lumen and to have a guide for further bouginage if necessary (see Chapter 6 for thread swallowing instructions).

Bouginage

Most esophageal strictures can be successfully treated by dilatation — especially Grades 1, 2, 3 and 4A (page 1[illegible]). As stated by Fatti *et al* (7), "grade 4B strictures are always difficult to manage but dilatation should be attempted until otherwise contraindicated." As long as progressively larger bougies can be passed, the prognosis is hopeful, but when repeated attempts fail to effect improvement, dilatation should be abandoned. Peroral dilatation using a previously swallowed thread as a guide has been successful in this clinic, which is mostly an adult clinic. In children the problem is sometimes more difficult. Thread swallowing may be accomplished in babies and children with the aid of candy or sugar-coated thread and persistent aid from experienced nurses. In babies once the thread is anchored in the intestine peroral bouginage may be attempted in the anesthetized patient, with or without esophagoscopy, but a thread must always be used as a guide. For repeated dilatations in children, however, it is advisable to avoid frequent anesthesia. It is preferable to perform gastrostomy, draw the thread out from the stomach with a tonsil pillar retractor and use retrograde bouginage, tying the bougie to the thread and pulling it upwards through the mouth after the method described by Tucker (2[illegible]).

If dilatation is carried out too rapidly hemorrhage perforation or mediastinitis may result. If these complications occur surgery must be considered but in these days of easy transfusions and antibiotics healing may take place without operation. The endoscopist who is treating stenosis by bouginage should appraise with the greatest care the type and degree of stenosis with which he is dealing and should consider surgical treatment for difficult problems before complications ensue.

Treatment of total atresia

In cases of total atresia of the esophagus a combination of peroral and retrograde esophagoscopy has been recommended We believe however that this method is very hazardous and that modern esophageal surgery is far safer and more satisfactory In some cases the total atresia is more apparent than real for although the patient may be unable to swallow water or his own saliva and x-ray examination may show a complete obstruction to the passage of thin barium a tiny lumen may be found and dilated at the time of esophagoscopy If however no lumen whatsoever is visible there are still two possible courses of treatment The first is gastrostomy, which will aid in maintaining the patient s fluid and nutritional balance, combined with retrograde and peroral esophagoscopy under fluoroscopic guidance Under even the very best conditions blind bouginage of a completely obliterated lumen The second method is surgery Since it has been shown by Sweet (20 21) and Nardi (16) that in suitable cases the stomach or colon can be brought up through the chest and anastomosed to the pharynx with good functional results this is the treatment of choice for most patients whose condition is not ameliorated by bouginage and probably for all patients with complete cicatricial atresia

Surgery

Surgical correction of the cicatricial and stenotic esophageal conditions resulting from caustic ingestion poses a technical problem of considerable magnitude The extensive inflammatory involvement (Fig 35) of both the length and the breadth of the organ renders excisional surgery difficult and hazardous Yudin (26) and Rienhoff (18) have summarized the development of current methods of restoring pharyngogastric or intestinal continuity Transthoracic esophageal resection with esophagogastrostomy is this method is difficult and dangerous since it is tantamount to currently *the most favored maneuver for surgical correction of*

caustic stricture of the esophagus We ourselves have had excellent results after carrying out a mediastinal colon transplant without esophageal resection (Fig 36), and we are inclined to favor the latter technic These two methods will be discussed in detail



Figure 35 Extensive caustic stenosis of entire subaortic thoracic esophagus This was of over [illegible]0 years duration and had not responded to bouginage

The surgeon should be aware that the ingestion of caustics frequently produces necrosis not only of the pyloric end of the stomach but also of the esophagus. The shortening of the stomach which is caused by such a burn may be a decisive factor in the choice of surgical procedure

As was earlier pointed out it is highly desirable to make the decision to operate before the patient's nutritional status requires gastrostomy Nevertheless, the procedure is unavoidable in many cases.

Esophageal resection with [illegible] [illegible]otomy

The periesophagitis and inflamm[illegible] [illegible]on which are this condition make [illegible] difficult and

ardous Particular care should be taken with hemostasis and resection should be carried out through normal tissue However, normal tissue is frequently difficult to define at the operating table The results of barium swallow do not necessarily constitute a reliable guide, since the inflammatory involvement is usually greater than is shown by the x-rays



FIGURE 36 Postoperative barium swallow of same patient (Fig 35) after substernal colon transplant Patient had complete relief of all symptoms

The surgical maneuvers involved are identical with those for esophageal resection in cases of carcinoma The reader is referred to Chapter 19 for details It is usually possible to mobilize the stomach adequately, even after gastrostomy for safe performance of esophagogastrostomy The stomach should be carefully inspected particularly in its pyloric portion for scarring and shortening Heineke-Mikulicz pyloroplasty is performed for any pyloric stenosis This procedure has the advantage of preserving enough length and flexibility so that the anastomosis is not jeopardized In some cases it may be necessary to bring the stomach up into the neck and carry out the anastomosis at this site Resection of the clavicle may facilitate surgery (20) but is not always necessary

(7) Esophagogastrostomy is usually inadvisable for patients with stomachs shortened by the combination of gastrostomy and pyloric stenosis

Colon substitution without esophageal resection

The use of large bowel as an esophageal substitute was established well before the turn of the century (26) Many surgeons have successfully utilized this organ during surgical treatment of esophageal carcinoma and atresia (3, 5, 9, 11, 13, 17) In 1951 Godard (8) attempted a two-stage subcutaneous prethoracic transplant of the ascending colon in a 7-year old child with esophageal lye stricture The patient succumbed to mediastinitis after the second stage In 1953 Montenegro *et al* (15) reported a successful two-stage transthoracic pharyngo-colo-jejunostomy in a 16 year-old boy with benign esophageal stricture

The development of carcinoma in the remainder of the stenosed esophagus is a theoretical objection to leaving this organ *in situ*, it is however extremely rare (see Carcinoma, page 223)

The technical hazards of attempts to remove a severely scarred esophagus certainly outweigh the theoretical desirability of this procedure An additional advantage of leaving the stenosed esophagus *in situ* is that the vagi are not interrupted and hence the distressing sequelae of gastric atony and retention are avoided

The use of colon rather than stomach has other advantages Since the colon can nearly always be extended well up into the neck without difficulty the upper anastomosis may be carried out wherever the surgeon thinks best, thus the operator is not forced to compromise at a lower site because the gastrostomized or scarred stomach may not be extended high enough In addition the colon may prove to have a higher resistance to the acid peptic secretions of the stomach Dragstedt and Vaughn (6) found that a loop of colon transplanted to the stomach of a dog was normal both grossly and microscopically after one year

The operation is performed through an abdominal and a cervical

incision (16) it can be carried out by two teams simultaneously A preoperative barium enema enables the surgeon to estimate how much colon is available and helps to rule out abnormalities of this organ Laparotomy is performed on the right side through a long

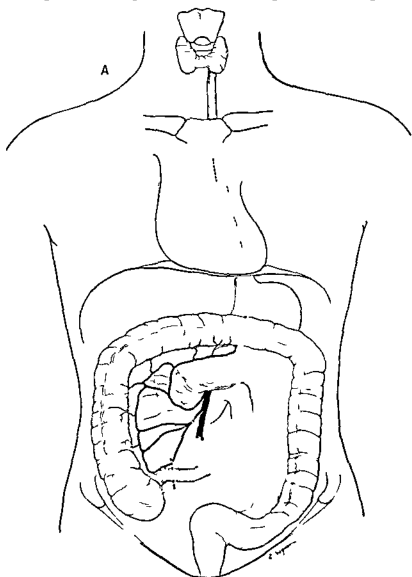


FIGURE 37 Colon substitution without esophageal resection
(A) Mobilization of the ascending and transverse colon (B) The cecum is passed through a substernal tunnel into the left side of the neck (C) Anastomoses (See pages 134 136)

vertical incision carried well up to the xiphoid The ascending and transverse colons are carefully mobilized retroperitoneally (Fig 37A) with every precaution taken not to injure the blood supply The arteries are dissected back to their origins, and rubber-covered bulldog clamps are applied to the right colic, ileocolic and other

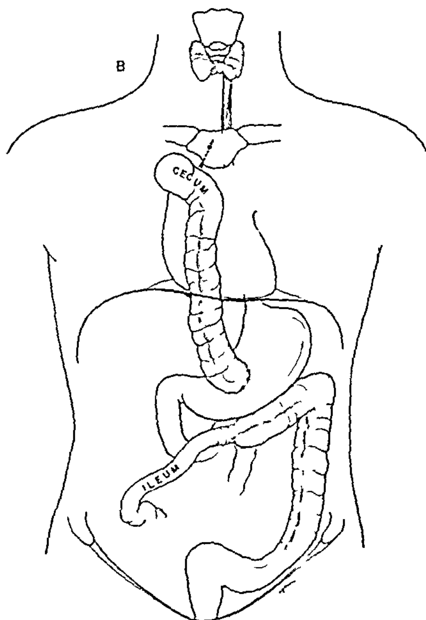


FIGURE 37 (*continued*) See legend page 133

arterial and venous branches to establish the adequacy of the mid-colic vessels after the ileum is transected Gastrointestinal continuity is restored by end to-end ileotransverse colostomy

Once the viability of the right colon has been established the blood vessels are divided at the site of application of the clamps Care is taken not to damage the arcades of the mesentery

The cervical incision is made parallel to the left sternocleidomastoid and the esophagus is identified and dissected free

A substernal tunnel is then created bimanually by blunt dissection as described by Robertson and Sarjeant (19) This route provides the shortest anatomical route and allows maximal utilization of colonic length It also permits operation without entering either pleural cavity

The segment of colon to be substituted is then passed *behind* the stomach and up through the tunnel into the neck (Fig 37B) Care should be taken not to rotate the segment since rotation will cause gangrene Passing the colon behind the stomach will allow the mid colic vessels of the vascular pedicle to pass innocuously and not press against the antrum and thus become a possible cause of gastric obstruction (Fig 37C) It should be emphasized however that this maneuver will not make available a greater length of transposed colon

The terminal portion of the ileum is then brought up into the neck and allowed to rest there

Cologastrostomy is then carried out on the anterior wall of the stomach in the position in which the segments lie best

The operator then returns to the neck and the viability of the colon is checked The appendix and any unused portions of colon and ileum are resected The cervical portion of the esophagus is transected and the distal end is closed with three layers of interrupted silk sutures and allowed to retract into the mediastinum A cervical esophagocolostomy completes the procedure (Fig 37C)

All incisions are closed without drainage Chest x rays are routinely made at the completion of the procedure to rule out the possibility that pneumothorax may have occurred undetected

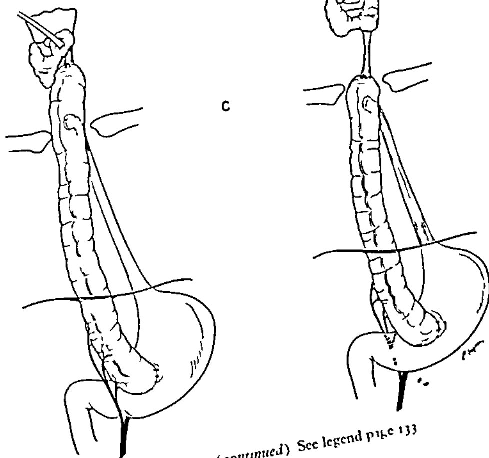


FIGURE 37 (*continued*). See legend page 133.

during dissection of the substernal tunnel. If pneumothorax is found, intercostal catheter drainage is instituted immediately.

PROGNOSIS

Nearly all cooperative patients can be successfully treated by one of the methods of bouginage outlined above. Although some patients enjoy complete restoration of function, others are obliged to live on a restricted diet. Surgery is the treatment of choice in patients with total atresia or in patients whose progress is unsatisfactory after proper bouginage has been attempted.

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Webs and Rings

13

A web is a membranous or partially fibrous structure which may extend across the lumen of the esophagus or may consist of a narrow band which encircles the lumen and partially constricts it. This pathologic entity, although occurring more commonly in the so called Plummer-Vinson (9) syndrome (anemia and esophageal web usually found in women), is not specific to that disease. Webs are not confined to the upper part of the esophagus but may be seen at all levels. They should not be confused with congenital atresia (Chapter 26).

Incidence

In 1935 MacMillan (8) reported that of 1600 patients with dysphagia examined in the out patient department of the Massachusetts General Hospital and the Massachusetts Eye and Ear Infirmary, 114 or 13 per cent had radiologically demonstrable esophageal webs. This lesion placed third in frequency to carcinoma and benign stenosis of the esophagus. Waldenstrom and Kjellberg (16), much to our surprise, describe this as the commonest esophageal disease in Sweden, twice as common as carcinoma. In a recent study Shamma'a and Benedict (1) analyzed 58 cases

of esophageal web. There were 51 females and 7 males. Forty-eight of the cases involved the upper esophagus; 25 of these were associated with anemia and fitted the description of the so-called Plummer-Vinson (9) syndrome. Twelve, or 25 per cent of upper esophageal webs (in 10 females and 2 males) showed no evidence of anemia. In 2 cases the webs were located in the middle sector of the esophagus. There were 4 cases *in our series of lower* esophageal webs or rings as described by Ingelfinger and Kramer (6), Schatzki and Gary (11), and Bugden and Delmonico (4). Four more cases of webs associated with benign mucous membrane pemphigus were also included in the report of Shamma'a and Benedict (12). They had previously been reported in detail by Benedict and Lever (3).

Etiology

In children and infants webs may occasionally represent a congenital malformation of the esophagus. In general, trauma of the esophagus, with possible esophagitis and ulceration, is the most likely cause of web formation. Contrary to the general impression, anemia is not an essential factor. In certain cases of lower esophageal ring there seems to be a definite web as described by Bugden and Delmonico (4). If such a web is present in the lower esophagus, the etiology may be based on trauma or esophagitis. By chilling the esophagus Respess (10) demonstrated a ringlike constriction in 7 of 31 patients in whom rings were not seen at routine fluoroscopy. Kramer (7) found that duodenal ulcer was associated with these rings. It seems likely that some of these lower esophageal rings may represent a definite structural entity that arises secondarily to inflammation or trauma, but a greater number are probably *a local segmental spastic phenomenon* of esophageal muscle. Some radiologists consider this to be a normal contraction phenomenon. The etiology of web seen in association with benign mucous membrane pemphigus is diffuse inflammation of the mucosa and submucosa which proceeds to scarring and adhesions. The

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Etiology

In children and infants webs may occasionally represent a congenital malformation of the esophagus In general trauma of the esophagus with possible esophagitis and ulceration, is the most likely cause of web formation Contrary to the general impression anemia is not an essential factor In certain cases of lower esophageal ring there seems to be a definite web as described by Bugden and Delmonico (4) If such a web is present in the lower esophagus the etiology may be based on trauma or esophagitis By chilling the esophagus Respess (10) demonstrated a ringlike constriction in 7 of 31 patients in whom rings were not seen at routine fluoroscopy Kramer (7) found that duodenal ulcer was associated with these rings It seems likely that some of these lower esophageal rings may represent a definite structural entity that arises secondarily to inflammation or trauma but a greater number are probably a local segmental spastic phenomenon of esophageal muscle Some radiologists consider this to be a normal contraction phenomenon The etiology of web seen in association with benign mucous membrane pemphigus is diffuse inflammation of the mucosa and submucosa which proceeds to scarring and adhesions The

process taking place in the esophagus in benign mucous membrane pemphigus is comparable with that affecting the conjunctivae The conjunctivae also show first diffuse inflammation with occasional bulla formation This gradually leads to the formation of adhesions which extend between the tarsal and bulbar conjunctivae of each lid as well as between the conjunctivae of the lower and upper lids These conjunctival adhesions are at first soft and pliable but with time they become fibrous and rigid

Associated Diseases

Carcinoma The association of the Plummer Vinson syndrome with carcinoma of the mouth the hypopharynx and the upper esophagus is well known and has been noted in numerous publications (1 12 13 16 17) In 1936 Ahlbom (1) found evidence of Plummer Vinson syndrome in 50 per cent of all women patients with carcinoma of the mouth and in 90 per cent of women with carcinoma of the hypopharynx and upper esophagus In Waldenstrom and Kjellberg s series of 26 patients (16) 5 had carcinoma of the esophagus and 3 carcinoma of the hypopharynx Welin (17) noted a higher incidence of carcinoma of the lower part of the hypopharynx in females He attributed this increase to the higher incidence of the Plummer-Vinson syndrome in women These and other authors regard this syndrome as a precancerous state the most probable mechanism being the atrophic changes in the mucous membrane of the mouth and esophagus which are associated with the anemia They advise iron therapy as a prophylaxis against the development of cancer

This interesting association is well illustrated in the Shamma a-Benedict series 9 patients (17 per cent) developed carcinoma 6 in the buccal cavity and 3 in the esophagus Five of the 6 patients with buccal carcinoma had a characteristic Plummer-Vinson syndrome Thus 1 out of every 5 patients with Plummer-Vinson syndrome in this series eventually developed carcinoma of the mouth All the cases were noted in women who complained of having had dyspha

gia from 8 to 40 years before the cancer became evident. There was no correlation between the severity of anemia and the incidence of carcinoma of the mouth. Two of the patients came from Prince Edward Island, Canada. One of them stated that 3 members of her family had died of carcinoma of the mouth and throat, and the other patient stated that 7 women in her immediate neighborhood had difficulty in swallowing. This is mentioned to reaffirm previous observations at this hospital of a high frequency of Plummer-Vinson syndrome and carcinoma of the mouth in women from Prince Edward Island. No satisfactory explanation has been offered.

None of the 12 patients who had upper esophageal web but no anemia developed carcinoma of the mouth. This significant finding suggests that anemia has a role in the pathogenesis of cancer of the buccal cavity. Three patients developed carcinoma of the esophagus: in 2 the tumor was in the upper esophagus and in 1 in the lower esophagus. The 2 cervical esophageal carcinomas developed at the sites of esophageal webs that had been demonstrated one to three years previously. Similarly, carcinoma of the lower esophagus developed at the site of a web that had been demonstrated ten years earlier.

Diverticulum. Diverticula of the esophagus were demonstrated in 3 patients. Two were in the cervical portion of the esophagus and were located above the web, and 1 was in the lower part of the esophagus. It is therefore important in operating upon a diverticulum to rule out an associated web (14) (see Chapter 16, page 181). If the web is unrecognized, results may be disappointing and the dysphagia for which the operation was performed may persist.

Thyroid disease. The finding of myxedema in 3 cases and of nodular goiters in 5 in Shamma'a and Benedict's report (12) prompted us to discuss this interesting observation here. A review of the literature reveals only a casual note on this association. Thus Plummer (9) stated that some patients whose dysphagia had been relieved and who were taking food freely developed a moderate hypothyroidism which necessitated the administration of thy-

roxin Six of Hoover's (5) 17 patients had nodular goiter No myxedema was found in these cases however

It would be tempting to speculate on the association of Plummer-Vinson syndrome and hypothyroidism Both conditions occur mainly in women and are associated with anemia The cutaneous thickening and hyperkeratosis of the face and hands which are frequently seen in myxedema are attributed to lack of thyroid hormone Histologic studies of the esophagus and the buccal cavity in Plummer-Vinson syndrome were reported to reveal thickened and hyperkeratotic epithelium (5 12) The similarity of these two histologic lesions warrants detailed histologic examination of the esophagus in myxedema

Pathology

Shamma a and Benedict (12) reported biopsies of 5 of the webs removed at esophagoscopy (Fig 38) These revealed a normal



Figure 38 Low power photomicrograph of a portion of an esophageal web which was resected from an area immediately above the cardia in a patient with typical x ray findings and with dysphagia There was an associated hiatus hernia.

squamous epithelial layer. The underlying connective tissue layer was the seat of a mild chronic inflammation with infiltration of mononuclear cells. Lymphoid hyperplasia with lymphoid nodule formation was noted in one instance. Smooth muscle was seen in two instances just beneath the connective tissue layer. Whether this smooth muscle was in the substance of the web or was situated in the wall of the esophagus is difficult to determine. No other abnormal findings were noted.*

The histologic appearance of the web is not specific. Degenerative and precancerous changes of the epithelium as noted by others were not observed in the Shamma'a-Benedict series. Hoover (5) examined an esophageal web that had been removed with punch forceps. It consisted of two layers of mucous membrane with a small amount of fibrous tissue between them. Suzman (13) reported hyperkeratinization of the epithelium with areas of atrophic degeneration in the esophagus.

Symptoms

Dysphagia is the one outstanding symptom; its duration may be a few months or many years. It is rather surprising and discouraging to learn that some patients who have difficulty in swallowing are told by physicians that they are hysterical (15) and are not even examined by x-ray or esophagoscopy — particularly when the latter may give immediate relief.† In addition to difficulty in swallowing there is sometimes pain on swallowing, especially when dry solid foods are ingested. If secondary anemia is present it should probably be regarded as due to dietary insufficiency, as also is true of the concomitant glossitis and fissures at the corner of the mouth. There may be exacerbations and remissions of the dysphagia, probably de-

* The authors wish to express their thanks to Dr Leonard Atkins of the Massachusetts General Hospital Department of Pathology for studying the histologic findings.

† We do not agree with Vinson's (15) concept of hysterical dysphagia. In fact, we believe such a diagnosis is almost never justifiable.

pending on the amount of infection and food irritation present. Cough may be a prominent symptom, caused by the overflowing of food into the trachea.

All of the 25 patients in the Massachusetts General Hospital (12) series had hypochromic anemia, with hemoglobin levels ranging from 3 to 11.8 gm. Blood smears made in 17 cases showed varying degrees of anisocytosis, poikilocytosis and hypochromia. No leukopenia was demonstrated. The serum iron level was low normal, 65 mcgm. per 100 ml. of serum, in the one patient in whom it was determined. The level subsequently rose to 150 with iron therapy, and there was concomitant improvement in the dysphagia. No adequate studies of gastric secretions were performed. The duration of symptoms in our series varied from as little as four months to as

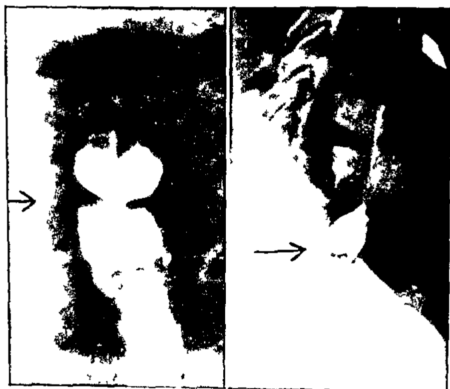


FIGURE 39. X-ray appearance of esophageal web. Left, anteroposterior view; right, lateral view.

PLATE IX Esophagoscopy

1 Typical fibrotic appearance of lye stricture of the esophagus as seen through the esophagoscope Bouginage was carried out with considerable relief The marked fibrosis which occurs in lye stricture makes it impossible for the lumen of the esophagus to return to normal size

2 Esophageal web as seen through the esophagoscope Note the very thin membranous appearance with small lumen on the left This web was readily divulsed by the esophagoscope and the patient was completely relieved

3 Esophageal varices as seen through the esophagoscope

4 Esophagoscopic appearance of benign leiomyoma shown by x ray in Figure 67 Instead of the carcinoma suggested by x ray esophagoscopy disclosed a smooth rounded lesion arising chiefly from the left wall of the esophagus Biopsy proved leiomyoma Bronchoscopy was negative

5 Smooth narrowing of the esophagus with very little gross evidence of malignancy Biopsy however revealed carcinoma The importance of biopsy in the differential diagnosis of benign stricture and carcinoma cannot be overestimated

6 Smooth narrowing of the esophagus with induration Biopsy showed carcinoma

7 Nodular irregular, polypoid mass typical of carcinoma as seen by esophagoscopy

8 Bronchoscopic appearance of esophageal carcinoma invading the left main bronchus near the carina – obviously inoperable

9 Endoscopic view showing penny in the esophagus of a six year old child – the penny was readily removed by esophagoscopy using rotation forceps

10 Endoscopic view of denture stuck in the esophagus just below the cricoid The denture was successfully removed by endoscopy

11 Sharp edge of flat triangular rather jagged chicken bone stuck in the upper end of the esophagus In spite of sharp points this was removed rather easily under local anesthesia using rotation forceps Excellent follow up results

12 Prune stone stuck at lower end of esophagus probably due to esophageal spasm secondary to a gastric ulcer as no other cause could be demonstrated by x ray or by esophagoscopy The stone was readily removed under local anesthesia using rotation forceps Since it was too large to bring into the lumen of the esophagoscope it was trailed at the tube mouth and removed with the esophagoscope The patient was relieved of his dysphagia

























Plate IX Esophagoscopy

incidental hiatus hernias, it is frequently of no clinical significance. Before such a ring is assumed to be causing symptoms a food-barium or barium filled gelatin capsule should be administered. If the dysphagia is coincidental with a holdup of the opaque test substance at the constriction, one may assume that the ring is causing the patient's symptoms. Sometimes esophageal webs are not demonstrated by x-ray examination, *therefore in every case of dysphagia,* even after a negative x-ray examination, esophagoscopy should be performed. See Figure 41 for x-ray appearance of web in pemphigus.

Esophagoscopy

Esophagoscopy demonstrates a gray membranous or fibrous structure usually located just below the cricopharyngeus (Plate IX, [illegible]). It looks like a diaphragm stretched across the esophagus and appears to leave practically no lumen. Bougies may be passed to stretch the web gradually, and finally the esophagoscope may completely divulse it. Very slight bleeding may occur. Although Bugden and Delmonico (4) and Bartlett ([illegible]) have demonstrated a definite web-like structure at the lower end of the esophagus *in* cases of esophageal ring such webs have not, so far, been demonstrable by esophagoscopy. If the pathologic basis of lower esophageal ring is really a web, it is very difficult to understand why such a web like structure has not been seen with the esophagoscope even under ideal conditions with a 53 cm. esophagoscope and general anesthesia being used for complete relaxation. The webs seen by esophagoscopy in cases of benign mucous membrane pemphigus may differ from other esophageal webs in having the appearance of fine thread like adhesions resembling cobwebs (Fig. 4) running vertically, obliquely and transversely.

SURGICAL TREATMENT

The indication for surgery is failure of endoscopic diagnosis and treatment. At the time of thoracotomy the web may be impossible



Figure 41 X ray appearance of multiple webs seen in benign mucous membrane pemphigus.

Figure 4 Unusual type of web seen in benign mucous membrane pemphigus.

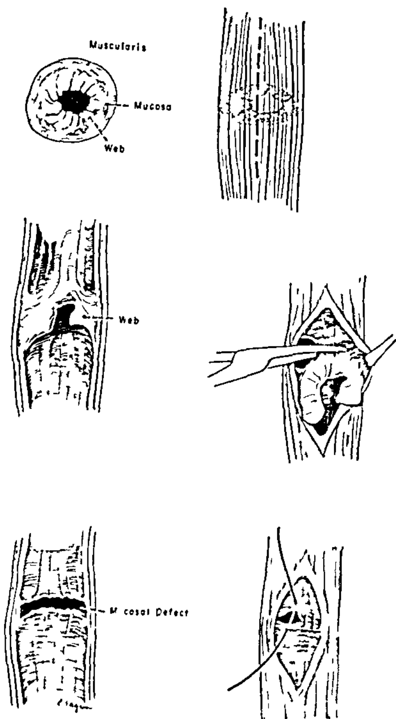


FIGURE 43 Transesophageal procedure for membranous esophageal web If the web is situated low near the cardia it may be approached from below through a gastrotomy

to palpate, therefore insertion of a nasogastric tube to the upper surface of the web helps the operator to localize by palpation of the tube the exact level of the web He can then determine where to make the esophageal incision A longitudinal incision is made over the web and the latter is excised with curved scissors The mucosal defect is repaired with fine interrupted silk sutures and the esophageal incision is closed longitudinally with the same type of suture (Fig 43) Results are uniformly good

Prognosis

The prognosis of esophageal web is excellent Usually one treatment by esophagoscopy with divulsion of the web is sufficient to give dramatic and permanent relief The prognosis of esophageal ring also is good if a web can be demonstrated If, however there is no web, the prognosis must be guarded

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Achalasia (Cardiospasm)—Megaesophagus

14

Achalasia of the esophagus is defined as failure of the lower esophagus to relax. It represents a mechanical obstruction of the passage of food from the esophagus to the stomach in the absence of an organic stricture. When this condition has been present for more than a few months it is frequently accompanied by dilatation of the esophagus — termed megaesophagus. To us 'cardiospasm' represents the same disease as achalasia; literally it signifies persistent greater-than normal tonic contraction. We believe that achalasia is a better term for this disease.

Incidence

As a cause of esophageal obstruction achalasia ranks next in frequency to carcinoma. It may occur in childhood or old age but is most common in middle life. Cases have been reported in the newborn (34). Its incidence is about equal in both sexes. Vinson (35) believes it to be the most frequent cause of dysphagia in women.

Etiology

It is generally agreed that the fundamental disorder in achalasia is lack of neuromuscular coordination of the esophagus

Fluoroscopic (33) and cineradiographic (5) studies of this condition have revealed absence of normal peristaltic waves The primary esophageal waves progress only to the level of the suprasternal notch and no secondary waves are seen The fluoroscopist frequently observes shallow, purposeless contractions with waves of antiperistalsis There is no coordinated relaxation of the lower esophagus at the cardia

Studies of intraluminal pressure have demonstrated that the entire esophagus is involved in the pathologic physiology of cardiospasm (4) Esophageal contractions are disorganized spontaneous and ineffective (32) To a physiologist, "patients with cardiospasm have a defect a defect which prevents them from initiating a standard primary peristaltic wave the true peristaltic wave This wave is the key which opens the door to the stomach the cardia Without this key the door must be forced so to speak (4)

Knight and Adamson (20 22), using cats, produced by means of vagal section, a condition similar to cardiospasm Ferguson (10) obtained the same results in monkeys In man cardiospasm has been reported as a complication of vagotomy performed for peptic ulcer (25 37)

Etzel (9) Rake (27) *and Lendrum* (23) *have* shown that in human beings with achalasia there is no abnormality of the vagus nerve They did however find a striking loss or complete absence of ganglionic cells (Auerbach's) from the myenteric plexi These plexi are believed to form relay stations for the vagal fibers on their route to the esophageal musculature Not only were the ganglia diminished or absent but in some cases there was evidence of active inflammation fibrosis and atrophy These changes were not limited to the narrowed segment at the cardia but were found equally often in the more proximal dilated portions of the esophagus *Whether these changes are the cause of* cardiospasm or merely

the effect of chronic distention has been debated for years The bulk of evidence indicates that the pathologic changes in the ganglion cells have a causal relation to the disease

It has been suggested by Etzel (9) that chronic vitamin B deficiency plays a part in the disease complex that includes megaesophagus megacolon and megaureter The therapeutic use of vitamin B however has not yielded favorable results

Jorge Broda, and Mealla (18) have reported one case in which achalasia and megaesophagus were associated with pellagra Ninety two per cent of Etzel s 28 Italian patients with megaesophagus and megacolon came from Veneto a district in which pellagra is common

PATHOLOGY

In early achalasia no gross lesion is found at the cardia As functional obstruction continues the proximal portion of the esophagus initially shows hypertrophy of both muscle layers (17) This stage of hypertrophy, with or without minimal esophageal dilatation has been termed the ' compensated stage of the condition (17 23 30) When pressure continues over a period of time decompensation occurs with dilatation and thinning of the muscular wall In long-standing cases the term megaesophagus is well suited to the lengthened, tortuous dilated gullet whose capacity has increased to fifteen or twenty times its original volume (Fig 44)

Naturally such an organized train of events does not occur in every case and patients may present themselves with a stabilized form of this disease process at any compensated or decompensated stage

Sloper (30) has reported 7 cases of diffuse muscular hypertrophy of the esophagus and has found 25 others reported in the literature He considers that this probably represents an early and fully compensated stage of achalasia

Sweet (31) on the basis of his clinical experience believes that there are two distinct types of achalasia The more frequent type

Type 1, comprising about 75 per cent of the cases, is that characterized by an enormous dilatation of the esophagus above the distal segment When exposed at operation the terminal segment just proximal to the cardia is amazingly small in diameter, with abnormally thin muscular layers making a striking contrast



FIGURE 44 Unusually high tortuosity in patient with marked achalasia and megaesophagus.

with the enormously dilated, thick-walled portion above In Type 2 achalasia approximately 25 per cent of the cases present an entirely different picture At operation the lower segment of the esophagus unlike that of the first group, in which it appears atrophic, is actually thickened though not dilated as it is above this level This appearance is due to the occurrence of a singular degree of hypertrophy of the circular muscle fibers in this portion " Sweet does not consider that one type of disorder evolves from the other Hawthorne *et al* (15), however, were unable to find any such correlation among symptoms, roentgen picture, and gross findings at operation and they are inclined to believe that these two extremes of deformity are related to the duration and the severity of the lesion rather than to its clinical type

Symptoms

Most patients report no difficulty in the act of swallowing but they complain that the food lies in the esophagus and does not reach the stomach They rarely note real vomiting — merely regurgitation or overflowing Some patients can empty the esophagus at will by regurgitation Nocturnal overflowing, a common complaint, is very troublesome, as it soils the pillow or the material is aspirated into the larynx with resultant cough In long standing cases nocturnal wakefulness owing to excessive cough is one of the most distressing symptoms Patients may eat well at some meals and not at others They may eat well when they are alone but become upset when in company Emotional strain or excitement before or during a meal often leads to an attack and under such circumstances patients may have to leave the table in order to empty the esophagus Such an embarrassing situation makes them avoid social events and public eating places Food may sometimes be forced into the stomach by drinking large amounts of fluids or by changing position or by holding the breath and bearing down Pain is seldom a prominent symptom although early in the disease associated esophagospasm may cause substernal discomfort The oc

currence of pain should lead the physician to question the diagnosis of achalasia

Complications

Pulmonary When food material is regurgitated and then aspirated into the tracheobronchial tree pulmonary complications are likely to develop These may include bronchitis, pneumonitis pneumonia, bronchiectasis and even lung abscess The pulmonary complication is sometimes so striking and so acute that the underlying achalasia is not even suspected Sudden death from asphyxia is rare but may occur Repeated bouts of aspiration may lead to pulmonary fibrosis (For a complete review of this subject the reader is referred to Breakey, Dotter, and Steinbert [3])

Carcinoma of the esophagus We have seen an occasional case of esophageal carcinoma which developed in a patient with achalasia of long duration Possibly prolonged irritation due to food stasis produces conditions which favor the development of carcinoma When carcinoma does develop in a patient with achalasia the diagnosis is difficult because the symptoms are already masked by esophageal obstruction and x-ray examination is likely to be inaccurate because of the presence of food material in the esophagus Esophageal carcinoma complicating achalasia has been the subject of several recent reports (6)

Carcinoma of the stomach Carcinoma of the stomach may occur in patients with achalasia (Fig 45) In such cases the carcinoma is likely to be overlooked not only because the patient already has gastrointestinal symptoms caused by his achalasia but also because at x ray examination too little barium passes through the cardia to permit satisfactory visualization of the stomach

Diagnosis

When a patient gives a history of long standing intermittent dysphagia one must think of achalasia as a possible cause If the

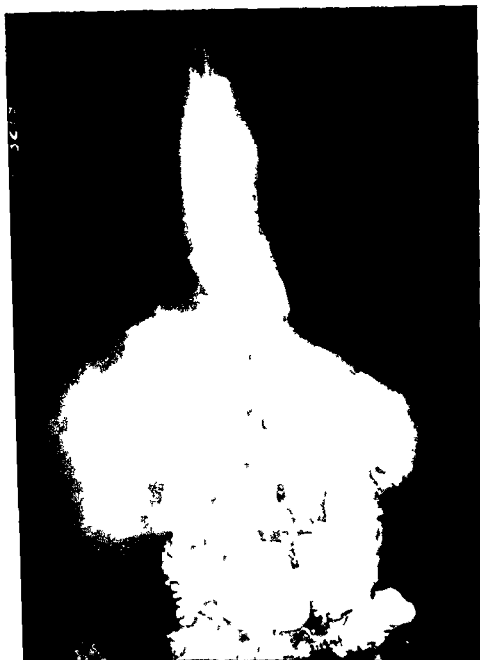


FIGURE 45. X ray showing achalasia and carcinoma of stomach. The latter was overlooked because the attention of the clinician and radiologist was focused entirely on achalasia.

attacks of dysphagia seem to be precipitated by emotional upsets, the diagnosis of achalasia is all the more likely. Physical examination usually is not helpful although it may possibly reveal widening of the mediastinum.

X-ray examination is the most important diagnostic method; it usually reveals a cone-like narrowing of the lower end of the esophagus with dilatation above it. A dilated and tortuous elongated esophagus is characteristic of megaesophagus and occurs only in achalasia. Sometimes the cardiac orifice relaxes well and permits barium to pass into the stomach; at other times the orifice remains firmly contracted or may respond to the inhalation of an amyl nitrite pearl. The gastric air bubble is usually absent.

Esophagoscopy. Since x-ray examination is not always 100 per cent conclusive in achalasia, and since it may be particularly difficult when food and secretions are retained in a dilated esophagus, it is advisable to recommend at least one diagnostic esophagoscopy in all cases of achalasia (Plate X).

In early cases in which obstruction is not marked, the esophagus may appear entirely normal and no change may be observed at the cardiac orifice. If slight obstruction is present, small amounts of retained food particles and liquid may be found in the esophagus. In long standing cases in which esophageal lavage has not been performed, the esophagoscopist encounters large amounts of retained food and secretions — sour, inspissated and stagnant material — some of which adheres to mucosa which is markedly reddened and edematous. The mucosa itself may bleed easily and may show small areas of erosion. Since such material cannot be adequately aspirated by esophagoscopy, the first such examination is unsatisfactory, and the procedure must be repeated after preliminary lavage with the large stomach pump and a warm weak sodium bicarbonate solution. Subsequent esophagoscopy should reveal a relatively clean esophagus; the mucosa, however, may still be somewhat red and edematous and may bleed easily. No organic obstruction is found at the cardiac orifice. Biopsy of the mucosa shows chronic esophagitis. In an elongated tortuous esophagus it may be difficult

to find the cardiac orifice In such cases the 53 cm esophagoscope should be used The esophagus may be so dilated tortuous and thrown into folds that one gets the impression he is looking into the stomach

Differential Diagnosis by Esophagoscopy

Peptic stenosis The differential diagnosis includes peptic stenosis which has sometimes been confused with achalasia at x-ray examination At esophagoscopy it is easy to differentiate achalasia and benign peptic stenosis since in achalasia the esophageal lumen is not narrowed or inflamed and there is no difficulty in passing an esophagoscope to the cardiac orifice There may however be some spasm at the orifice, and passage through it is often best accomplished by using the long esophagoscope under general anesthesia

Peptic ulcer of the esophagus may be confused with achalasia in a few rather unusual instances However the differential diagnosis is easily made at esophagoscopy In ulcer there is either a rather marked inflammatory narrowing above the ulcer or else the ulcer can be seen with the esophagoscope In achalasia as previously noted the esophagoscope passes readily to the cardiac orifice and usually through it into the stomach

Carcinoma of the esophagus Since patients with achalasia already have symptoms of esophageal obstruction and since x-ray examination may be difficult because of retained food and secretions in a dilated esophagus esophagoscopy should be performed to rule out the possibility of carcinoma Usually carcinoma superimposed on achalasia can be seen by esophagoscopy and the diagnosis established by biopsy However in the rare case in which a very small carcinoma has developed in achalasia and megaesophagus the tumor may be missed by x ray as well as by esophagoscopy Therefore in such a case cytologic study is important

Carcinoma of the stomach also must be considered in cases of achalasia with unrelieved obstruction We know of at least two cases of achalasia in which diagnostic attention was focused on the

esophagus and cardia but carcinoma of the stomach developed later without being recognized. Careful x-ray examination and long open-tube esophagogastroscopy as well as flexible-tube gastroscopy are important in the study of such cases.

Cancerphobia. Many patients with esophageal obstruction immediately suspect that they have carcinoma, and although they can be somewhat reassured by a negative x-ray examination, further confirmation of a normal esophagus by esophagoscopy helps greatly to convince them that no carcinoma is present.

TREATMENT

Diet

There is no universal rule for diet in achalasia. Most patients do not tolerate ice-cold foods or drinks. Some patients do better with solids than with liquids, and many patients can wash down solid food with liquids. Fried or highly seasoned foods should be avoided. Salad should be avoided, as parts of it are likely to remain in the esophagus. Alcohol is undesirable, and smoking should be eliminated. Drug therapy is usually ineffective. Antispasmodics are rarely helpful, the most useful being nitroglycerin in tablet form 1/100 grain (0.3 mg.) held under the tongue just before each meal. *In nervous persons small amounts of barbiturates may be indicated.* Atropine sulfate apparently does not help. The nitrites which relax smooth muscle, intact or denervated, may initially be effective, but tolerance develops rapidly (28, 29).

Bouginage

The passage of a rubber covered mercury filled bougie (see Figure 15) is definitely helpful. Sometimes one or two treatments seem to relieve all symptoms completely *for many years* (Fig. 46). On the other hand, some patients require bouginage daily, and in such cases the patient may purchase his own bougie and use it at home. We have one patient who passes the bougie every morning



FIGURE 46 Typical x ray appearance of achalasia with megaesophagus. Marked relief by bouginage

before breakfast and even took it with her on a recent trip to Europe In spite of the fact that she was offered surgical therapy she prefers daily bouginage After a few treatments some patients feel so well and eat so normally that they no longer return to the office or out patient department A few patients however hate the

bougie and receive little or no benefit from its use Number 50 French is an average-sized bougie It may be passed blindly with impunity because it is blunt and rounded at the end No other type of bougie should be passed blindly, because of the danger of perforation Bouginage with the mercury bougie is an office procedure carried out with the patient fasting, sitting upright in a chair with head flexed and chin on chest In this position the cricopharyngeus is relaxed and the bougie usually passes quite easily There may be delay, discomfort, or pain in the subxiphoid region as the bougie reaches and passes the cardiac orifice Usually no preliminary sedation or cocainization is necessary If this type of bouginage is not satisfactory, some type of dilating bag may be used Good results are often obtained with the Browne-McHardy bougie This is a mercury bougie with a dilating bag attachment (see Figure 16) An ordinary #50 French mercury bougie dilates the cardiac orifice to a diameter of approximately 1 7 cm With the dilating bag attachment the orifice is stretched to 4 or 5 cm In some cases of tortuous megaesophagus the bougie will not enter the cardiac orifice at such times fluoroscopic guidance may be helpful If this fails esophagoscopy under general anesthesia using the 53 cm esophagoscope and bouginage under direct vision with the tip of the esophagoscope being passed into the upper stomach may be helpful as a method of stretching the cardia When medical treatment and bouginage fail surgery should be advised

According to Franklin (12) Thomas Willis in his *Pharmaceutica rationalis* (1674) describes a case in the following words

A strong man and otherwise healthful enough labouring for a long time with often vomiting he was wont very often though not always presently to cast up whatsoever he had eaten At length the disease having overcome all remedies he was brought into that condition that growing hungry he would eat until the oesophagus was filled up to the throat in the meantime nothing sliding down into the ventricles he cast up raw (or crude) whatsoever he had taken in when that no medicines could help and he languished away for hunger and every day was in danger of death I prepared an instrument for him like a rod of a whale bone with a little round button of sponge fixed to the top of

it, the sick man having taken down meat and drink into his throat presently putting this down in the oesophagus he did thrust down into the ventricle its orifice having opened the food which otherwise would have come back again and by this means he hath daily taken his sustenance for fifteen years and doth yet use the same machine and is yet alive and well who would otherwise perish for want of food

Surgery

The finding that in animals sympathectomy reverses or prevents the development of the achalasia which follows vagotomy in combination with the fact that myenteric ganglion cells are absent in clinical specimens would suggest sympathectomy as a logical surgical maneuver However the reported results of this procedure (21, 24 26) have not been impressive, and it is currently agreed that direct surgical procedures involving the esophagus itself are the most desirable The most effective and widely used procedures are cardiomyotomy and cardioplasty

Cardiomyotomy

This procedure (Fig 47) is a modification of that described by Heller (16) and originally suggested by Gottstein (14) It is essentially an extramucosal esophagocardiomyotomy We prefer the transthoracic approach but the maneuver can be effected equally well through the abdomen

A nasogastric tube is inserted into the stomach before operation Through the bed of the eighth rib or eighth intercostal space the lung is reflected upwards and the mediastinal pleura incised The esophagus and the cardia are mobilized Soft rubber wicking is passed behind these for traction There is usually little inflammatory reaction and dissection along proper cleavage planes is not difficult A longitudinal area for incision is chosen and any vessels in this sector are carefully clipped or stitch ligated The muscularis is then carefully incised longitudinally and the incision carried down to the mucous membrane of esophagus and stomach

Care should be taken not to perforate the mucosa, this can easily happen, particularly in the stomach Any perforations should be carefully repaired with interrupted sutures of fine silk

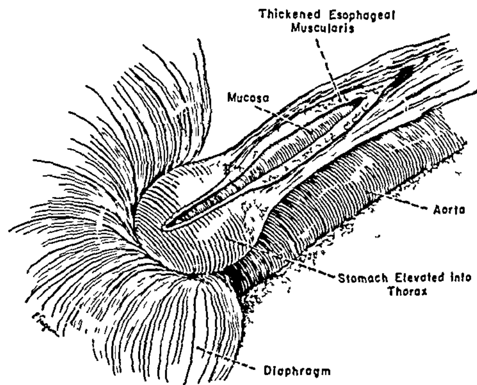


FIGURE 47 The modified Heller procedure All muscle fibers should be completely divided throughout the length of the incision Note that the incision extends on to the stomach

We believe that both the depth and the length of incision are of considerable importance Care should be taken that all the muscle fibers are thoroughly divided throughout the length of the incision The incision itself is so placed that two thirds of its length is in the esophagus and the remaining third on the stomach It should be at least 10 cm in length and in cases of extensive hypertrophy it should extend the length of the hypertrophic involvement

Wangensteen (36) has emphasized the importance of complete rupture of the muscle fibers in the Heller procedure He supple

ments the operation with air inflation by a Foley balloon type catheter under direct vision at the time of surgery

The same procedure can be carried out through the abdomen — after the left lobe of the liver is mobilized by dividing the left coronary ligament Increased frequency of pyloric hypertrophy has been noted in patients with cardiospasm (30) and where this is suspected or where there is any history of gastric retention or delayed emptying the abdominal approach with concomitant pyloroplasty is advisable

The nasogastric tube is left on suction for 24 to 48 hours The patient s diet is gradually increased to normal over a period of a week

There is little doubt that this procedure is the safest and most effective available at the present time

Cardioplasty

In the occasional case in which the Heller procedure cannot effectively be executed owing to the thinness of the terminal narrowed segment a cardioplasty may be indicated (see Chapter 11, page 115, for technic of this procedure)

Though a cardioplasty almost always provides excellent immediate symptomatic relief long term follow-up of these procedures by many competent observers has been almost uniformly unsatisfactory (1 . 7 8 11 13) The destruction of cardial competence by this procedure results in a high incidence of reflux peptic esophagitis with symptoms and complications that are frequently more serious and distressing than the original disease Kay (19) reported 9 of 3 patients having a cardioplasty had evidence of esophageal bleeding and anemia In 4 the bleeding was severe

Nevertheless the surgeon may frequently have no choice but to carry out such a procedure Should the patient fall into an older age group the chances are good that no complication will ensue, since this older patient usually has hypo- or achlorhydria (15) Particular care should be taken to reduce the operative site below

the diaphragm as in repair of hiatus hernia This maneuver may be of help in avoiding regurgitation and esophagitis

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ESOPHAGOSPASM (CURLING)

15

Esophagospasm (curling) is diffuse spasm of a long segment of the esophagus usually involving the lower half Although it is a well-recognized entity it is definitely rare Other terms falling in this category include pseudo diverticulosis bladder spasms segmental spasms reflex spasms tertiary spasms spastic stricture rosary esophagus and corkscrew esophagus Achalasia or so called cardiospasm is probably a different disease entity but Van Laeer and Keet (14) Templeton and Moore (12 13) Levrier and Bouloux s (9) Hillemand *et al* (5 6) and Chene and Poirier (.) have noted that these appearances may be seen in achalasia

One of the earliest reports is that of Moersch and Camp (10) who reported 8 cases characterized by intermittent dysphagia and substernal pain Schmidt (11) reported 17 cases of diffuse spasm of the esophagus in 15 of whom this condition was the primary chief complaint Eskridge and Peake (3) reported 19 cases of curling of the esophagus They state in the average case that is symptomatic but not completely obstructed one sees a most unusual sight under the fluoroscope The patient swallows the barium and when the bolus fills the lower half of the esophagus there suddenly appear varying numbers of ring shaped constrictions that

may completely occlude the lumen and do not move up or down This is usually before the first wave of peristalsis, and when that arrives the portion above the tight constriction distends until some of the barium is regurgitated above the peristaltic wave In some patients the pressure is so great that diverticula are seen to protrude and become permanent They are certainly not traction or pulsion diverticula but are secondary to the pressure that must cause actual mucosal and submucosal herniation through the muscle coats The peristaltic wave is entirely independent of the constricted areas and passes on down to the cardia Then after a few seconds the constrictions gradually relax and the esophagus distends to a larger than average size

The average age was 66⅓ The youngest patient was 49 years and the oldest 84 years Of the 18 cases, 7 or 39 per cent were symptomatic Of these 1 had mild 4 moderate and 2 marked symptoms Eleven were over 65 years of age and 9 over 70 There were 8 men and 10 women Complicating disease was common and since our contact with the patients was rather brief, we are sure that other conditions were probably present in most cases Those known to us are as follows cardiac disease 7, hiatus hernia 5 peptic ulcer 5, mild cardiospasm 3 and a number of others We are unable to say just how much the condition is influenced by the mental state One patient had an anxiety state and another was a chronic alcoholic The presence of complicating organic disease and old age predominates

Etiology

The cause of this condition is obscure, and many etiologic theories have been advanced Carlson *et al* (1) demonstrated in animals that spasm of the distal end of the esophagus can be caused by pharyngeal stimulation stimulation of the sciatic nerve traction on the vagus nerves sudden stretching of the stomach strong distention of the urinary bladder or large or small intestine and various types of mechanical stimulation of the gallbladder common bile

duct, and skin Numerous authors have noted that curling is related to various diseases such as gastric ulcer or carcinoma hiatus hernia duodenal ulcer coronary disease, gallbladder disease Parkinsonism, and so on Most authors agree that it typically occurs in nervous high-strung persons over the age of 50 who in most cases have some other disease also We have seen it both as a primary condition and also as one secondary to carcinoma of the stomach

Evans (4) has discussed the relation between esophageal contraction and cardiac pain He finds what he calls esophageal arrhythmia which is characterized by a spasm which affects the lower third of the gullet in many patients who have no heart disease but do have the type of pain that is found in cardiac disorders In conclusion he states that ' when the electrocardiogram is strictly normal in a patient with cardiac-like pain as it often is we shall gain help in our search for the cause of the pain if we look for the radiological sign of oesophageal arrhythmia

Van Wezel (15) reported an unusual case of esophageal spasm due to cold When barium was given at body temperature there was no evidence of spasm However when the same barium was suspended in a cold solution it produced intense spasm in two locations — first at the aortic knob and second about one inch above the cardia These spasms lasted as long as 5 to 10 minutes There was no evidence of any physical allergy or other systemic change the spasm was strictly a local phenomenon

Ismay (7) has reported a case of painful esophageal spasm which he otherwise terms corkscrew esophagus or curling When his patient swallowed barium the esophagus below the level of the aortic arch was seen to be repeatedly broken up by violent circular contractions — the corkscrew effect These contractions appeared very frequently and became more obvious when the patient lay flat They were associated with severe pains in the chest Between the attacks of spasm the esophagus appeared normal Atropine sulfate gave considerable relief probably by inhibiting the vagi and their action on smooth muscle

Lerche (8) credits Levner with describing spiral fibers in the

inner muscle layer of the esophagus is responsible for the tertiary contractions and curling

Levrier and Bouloujs (9) reported 4 cases of what they termed tertiary contractions of the esophagus In one case there were contractions of the lower third of the esophagus associated with hiatus hernia, in another, there was a corkscrew type of contraction in a patient who had had cholecystectomy for gallstones A third case demonstrated a pseudo-diverticulum type in a patient with a chronic gastric ulcer In the fourth case there was a mixed type showing tertiary corkscrew effect and pseudo-diverticulum in a patient with hiatus hernia These authors regard the condition as a neuromotor disequilibrium associated with old age

Symptoms

Dysphagia and pain are the characteristic symptoms The dysphagia is intermittent and there may sometimes be complete stoppage of food Regurgitation is likely to occur The lower substernal pain may awaken the patient from sleep, relief may be obtained by getting out of bed and walking around

At first the patients have no trouble with *solid food* but later even liquids may cause difficulty The dysphagia is likely to be *much more marked when the patient is excited or nervous* and at such times he may be unable to swallow water

Diagnosis

The diagnosis depends almost entirely on x ray examination which reveals the type of diffuse spasm shown in Figure 48 Esophagoscopy reveals a normal appearing esophagus with no definite area of obstruction

Treatment

The condition is so rare that no one standard method of treatment has been determined The use of *nitroglycerin drs under the*

tongue, as in achalasia should be tried Nervous excitement and fatigue should be avoided Patients should be instructed to eat slowly Esophagoscopy and bouginage may give temporary relief, and further relief may be obtained by the use of the Hurst mercury filled bougie as in achalasia



FIGURE 48 X ray picture of moderate esophagospasm (Courtesy of Dr Lewis Kane)

Surgical treatment The Heller procedure of esophagocardiomyotomy may be performed on the lowermost segment of the esophagus if there is evidence of hypertrophy of the muscle fibers in that area Any associated condition such as hiatus hernia should be corrected at the same time

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Diverticulum

16

An esophageal diverticulum is an outpocketing of the esophageal lumen with resultant formation of a pouch or sac.

Incidence

Diverticula are statistically more common in the thorax than in the neck. However, the type most commonly observed at clinical examination is the pharyngoesophageal diverticulum. This is probably due to the fact that the confining fascial planes of the neck produce earlier symptoms. The condition is about three times as frequent in men as in women, and most patients are well over 50 years of age when first seen.

Etiology

Esophageal diverticula are classified as either pulsion or traction diverticula (Plate XI) according to their supposed mechanism of formation.

Pulsion diverticula. In the pulsion type the intraluminal pressure of swallowing is considered the causative force. A diverticulum

may be the result rather than the cause of dysphagia. Incoordination between pharyngeal constrictors and relaxation of the cricopharyngeus may result in local hypertonia with resultant obstruction of swallowing (5). A spastic obstruction of this type or an organic one such as may be associated with a web when coupled with a weakness in the esophageal wall results in a diverticular bulge.

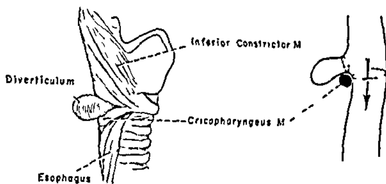
Pulsion diverticula are found at the upper (pharyngoesophageal) and lower ends of the esophagus. Those that occur at the lower end are usually situated just above the level of the diaphragm; they are termed epiphrenic. These diverticula are anatomically false diverticula since they consist of herniation of the mucosa and submucosa through a defect in the muscular wall.

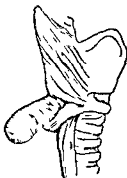
Traction diverticula. These are usually situated in the midesophagus in the subcarinal region of the mediastinum. They are produced by the pull upon the esophageal wall by the cicatrization caused by an adjacent inflammatory process — usually tuberculous lymph nodes. The diverticulum usually consists in a distortion of the esophageal wall. A small sacculation may form which is higher than the mouth of the sac. These are true diverticula in the sense that their walls contain all layers of the esophagus.

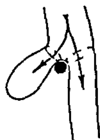
Traction diverticula rarely produce symptoms since they are broad necked and the sac is not dependent. They may be multiple and are usually found incidentally at x-ray. In rare cases a diseased lymph node may ulcerate into the lumen of the esophagus and trachea and produce an esophagotracheal fistula. (This problem will be considered in Chapter 6.) In addition, local infection may produce bleeding or an abscess.

Symptoms

Diverticula probably are present for many years before they produce symptoms. The commonest symptoms in order of frequency are dysphagia, regurgitation, gurgling noises in the neck, nocturnal choking and coughing, and weight loss. Symptoms may frequently be correlated with size and attendant distortion of the







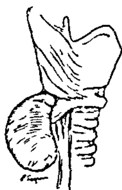




FIGURE 49 Development of esophageal diverticulum

esophageal lumen as the diverticulum enlarges (Fig 49) Epiphrenic diverticula are more likely to cause pain and dysphagia (6)

Complications

When there is much regurgitation and aspiration into the trachea pulmonary complications may occur Belcher (1) described 5 patients with pulmonary complications associated with esophageal diverticulum, 3 had developed lung abscess 1 pneumonia and 1 lipoid pneumonia Foreign bodies may lodge in a diverticulum and perforate causing mediastinitis As previously mentioned local disease processes may involve both trachea and esophagus and result in tracheoesophageal fistula Carcinoma in a diverticulum has been reported (Fig 50) but this is uncommon Hemorrhage is rare, but has occurred in 1 of our patients (2)

Diagnosis

Diverticulum should be suspected when any of the above mentioned symptoms are noted The diagnosis can readily be made by x ray examination (Fig 51A B) The diverticulum should be studied by fluoroscopy care should be taken to determine the size of the mouth and whether or not it empties readily In particular the adjacent areas of the esophagus should be examined for the presence of a web a tumor or any other possible cause of obstruction and dysphagia

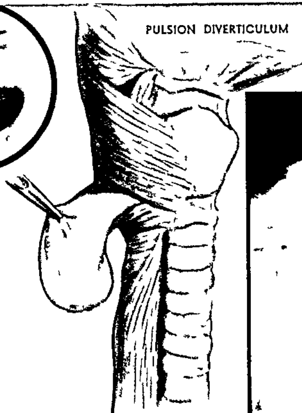
Esophagoscopy

Esophagoscopy is indicated only in certain types of diverticulum under certain conditions When the mouth of the diverticulum is wide and there is no obstruction of the esophagus below the diverticulum esophagoscopy is usually noncontributory On the other hand esophagoscopy may reveal esophagitis near the mouth of the diverticulum or may disclose an inflammatory stenosis be-



FIGURE 50 X ray appearance of carcinoma in a diverticulum of the esophagus The patient was a 60-year-old man with a 12 year history of regurgitation (Courtesy of Dr Richard H Sweet)









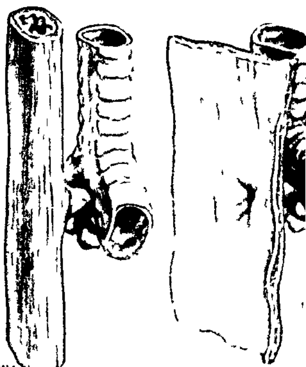


Plate XI. Diverticula of the esophagus (Courtesy Fletcher D. Woodward, M.D., Clinical Symposia 8:3 [illegible])

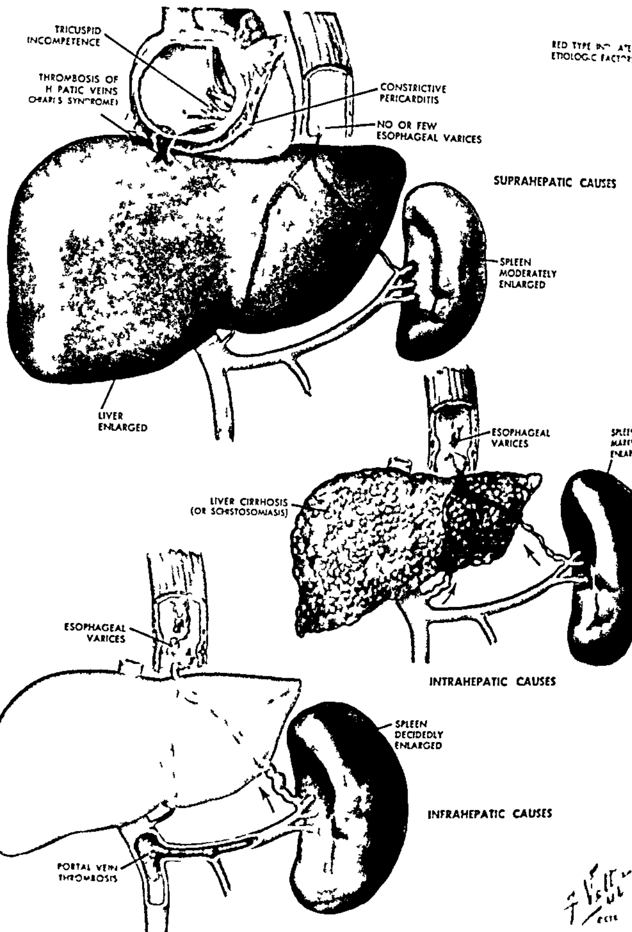


Plate XII Causes of portal hypertension (Courtesy The CIBA Collection of Medical Illustrations Volume 3 Part III)

low the diverticulum which can be benefited by bouginage or, in the case of web, by bouginage or divulsion. Habein *et al* (3) advises routine preoperative dilatation. On rare occasions moderately severe hemorrhage has originated in a diverticulum ([illegible]). In such a case the origin of the bleeding may be exactly determined by esophagoscopy.



FIGURE 51A. X-ray appearance of hypopharyngeal diverticulum arising from the posterior wall of the esophagus. Note the pressure defect on the esophagus produced by the diverticulum. There was an 8 hour residue within the diverticulum. The patient was a 53 year old man who had had slight dysphagia for several years and who had the feeling after swallowing of food remaining in the throat.

TREATMENT

The small asymptomatic diverticulum requires no treatment. The rare patient with minimal symptoms and an associated web or area of spasm below a small diverticulum may profit by preoperative dilatation.

Pharyngoesophageal diverticulum. The symptomatic diverticulum requires surgical extirpation. Von Bergmann (8) is credited



FIGURE 51B X ray appearance of diverticulum—mid esophagus—which was an incidental finding in a patient with duodenal ulcer The diverticulum was rigid and may have resulted from an old tuberculosis gland which adhered to the esophagus and later extended itself into the esophagus.

with the first successful removal of an esophageal diverticulum Early experiences with excisional surgery were associated with a high incidence of suture-line breakdown sepsis, and pneumonia and with a high mortality rate As a result, a two stage procedure was advocated for many years by Lahey and Warren (4) At the first stage the sac is dissected free and tacked up so that it may empty and not cause obstruction It is excised at a second stage with the surrounding adhesions serving as a prophylaxis against mediastinitis

We have not utilized the staged procedure, we consider that it has no advantages and presents disadvantages to both operator and patient as compared with a one stage procedure We do believe however, that in the very-poor risk emaciated elderly patient with obstruction, the first stage of the two stage procedure carried out under local anesthesia may serve as a definitive procedure and may occasionally salvage an impossible situation

The operative technic is patterned after that of Sweet (7) The incision is usually made along the anterior border of the left sternocleidomastoid and the esophagus exposed as described on page 60 The approach is usually made on the left since most diverticula are present on this side and since right sided diverticuli can be pulled over to this side without undue difficulty

The esophageal sac is carefully dissected free of adjacent structures particular care being taken not to injure the recurrent laryngeal nerves The junction of the neck of the sac and the esophagus is defined and the neck circumcised by an incision down to the mucosa (Fig 52) Stay sutures are then placed in the esophageal muscularis and the mucosa is divided as the sutures are inserted The mucosal sutures are placed so that the knots lie within the lumen

In the illustrations an excessive length of stump is shown intentionally for the purpose of clarity The mucosal neck should actually be transected almost flush with the normal esophageal lumen Care should be taken however that sufficient cuff is left so that the sutures will not compromise the lumen Finally the muscularis is reapproximated with fine interrupted silk sutures

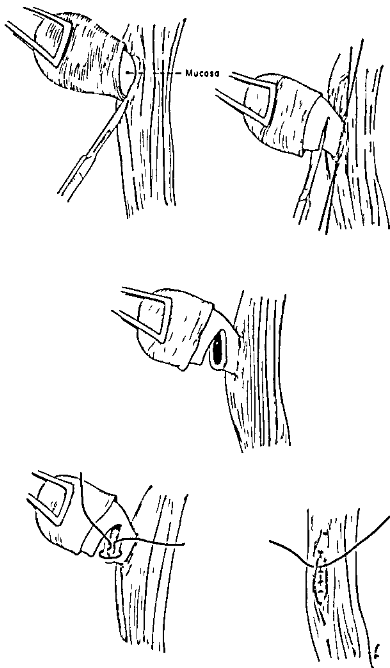


FIGURE 52 Steps in excision of esophageal diverticulum.

Before closing the esophageal lumen the surgeon should satisfy himself that there is no web or other obstruction in the esophagus below the site of excision If such is encountered it is dealt with accordingly A Levin tube is not inserted preoperatively or postoperatively The wound is closed without drainage Penicillin is administered parenterally for about 5 days After operation the diet is slowly advanced so that the patient is taking soft solids after a week

Epiphrenic diverticula These are most commonly found on the right side and accordingly are approached through a conventional right thoracic incision (Actually they may be approached from either side) Incision is made through the bed of the eighth rib or eighth intercostal space The technic of removal is identical to that of pharyngoesophageal diverticulum and the same precautions concerning a possible lower obstruction should be observed before the esophageal defect is closed

Prognosis

The results of the procedure are generally excellent and recurrence is unusual

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VARICES

17

Esophageal varices are dilated veins in the wall of the esophagus.

INCIDENCE

Esophageal varices almost always represent collateral channels that have developed secondarily to portal bed block. They are much more commonly found in men than in women.

ETIOLOGY

A discussion of the etiology of esophageal varices necessarily results in a review of the causes of portal hypertension.

Ekman (20) demonstrated collateral variceal circulation in all of 54 patients with portal hypertension and in none of 16 controls. He interprets his findings as strong indirect evidence that such collaterals are a manifestation of an increased pressure gradient between the portal and caval venous systems. When splanchnic venous outflow is obstructed, an increase in splanchnic pressure develops which is referred to as portal hypertension. This obstructing hypertension results in the development of a collateral circula-

tion Esophageal varices represent one set of such collaterals They are of clinical importance since they often rupture and bleed The maximum normal pressure in the portal vein is approximately 20 cm of water In portal hypertension it may rise as high as 50 cm or more This hypertension is a consequence of a variety of disease processes and is not a disease *per se* In 1945 Whipple (60) proposed that patients with portal hypertension should be divided into two groups those in whom the block is due to intrahepatic disease and those in whom the block lies outside the liver The latter group may be subdivided into supra- and infrahepatic block (Plate XII)

In the United States most patients fall in the first category, the block resulting from cirrhosis of the liver Patek (4[illegible]) found elevations of portal pressure in about 33 per cent of patients with Laennec's cirrhosis Ahrens (2) encountered portal hypertension in 50 per cent of patients with biliary cirrhosis Other causes of intrahepatic block include schistosomiasis sarcoid, hemochromatosis and metastatic carcinoma (47) In children hepatic agenesis or the so called *Cruveilhier-Baumgarten syndrome* (3) is sometimes seen In this condition the liver atrophy is thought to result from a congenital communication between portal and systemic circulation through a patent umbilical vein However Jahnke *et al* ([illegible]5) on the basis of personal experience are inclined to believe that the syndrome may represent a stage in the life history of portal hypertension regardless of cause in which the umbilical collateral circulation is excessively employed

Suprahepatic block (Plate XII) may result from heart failure hepatic vein occlusion or pulmonary fibrosis It is easily recognized since there is associated generalized venous hypertension Clinically bleeding varices are almost never a problem An exception to this is the Budd Chiari syndrome (11) which is characterized by ascites and esophagogastric anastomoses which develop secondarily to intrahepatic venous thrombosis

Infrahepatic block (Plate XII) is most commonly encountered in children and young adults Ekman ([illegible]0) found extrahepatic

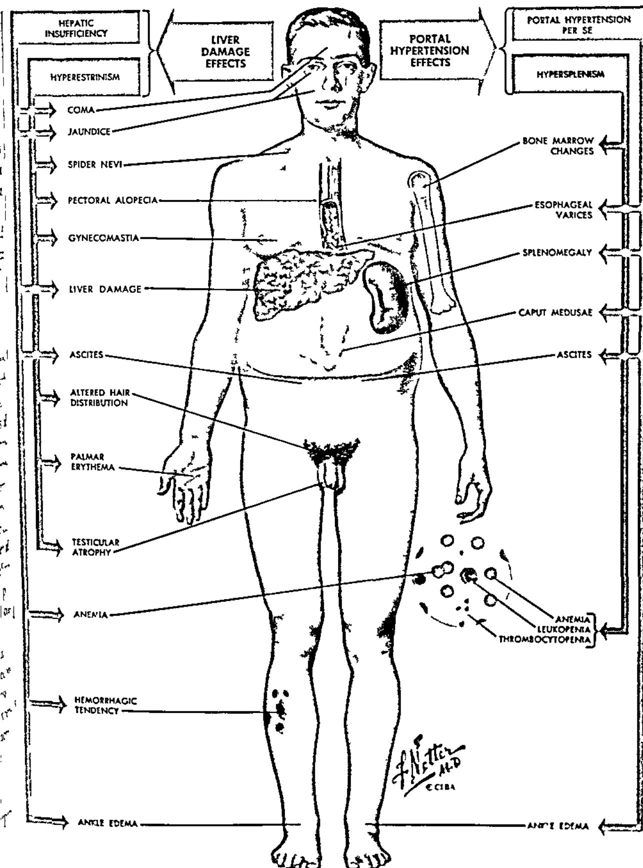


PLATE XIII. Clinical manifestations of cirrhosis. (Courtesy The CIBA Collection of Medical Illustrations, Volume 3, Part III.)





Plate XIV. Esophageal varices (Courtesy Fletcher D. Woodward, M.D., Clinical Symposia 8:3, 1956.)

block in [illegible]5 of 33 patients under the age of [illegible]1, whereas in a group of patients between 41 and 60 years of age who had portal hypertension 2[illegible] of 28 had intrahepatic block. Portal hypertension may result from thrombosis of the portal, superior mesenteric or splenic veins, from stenosis or atresia of the portal vein, and from cavernomatous transformation of the portal vein. A septic thrombotic process is believed to be the most common cause of sub hepatic obstruction. Efskind (19) showed that umbilical infection during neonatal life can spread along the umbilical vein and result in portal-vein thrombosis. Cavernomatous transformation of the portal vein is currently believed to represent a recanalized thrombus ([illegible]2, 60) rather than an angioma or a primary malformation. In adults subhepatic portal hypertension is usually secondary to tumor invasion or compression. Acute inflammation of the pancreas has led to the development of esophageal varices and hemorrhage (53).

Pathology

Esophageal varices are usually found in the lower portion of the esophagus, but in severe cases they may extend as high as the upper third. They frequently extend into the stomach, and not uncommonly hemorrhage may occur from dilated veins in the fundus of the stomach. Chiles *et al* (1[illegible]), in a study of 60 cases of ruptured esophageal varices, found that rupture had been caused by increased hydrostatic pressure in 39 per cent and by ulceration in 50 per cent. In the majority of cases of hemorrhage ulceration appeared to be a contributory factor in the rupture. Palmer and Brick (39), who measured varix pressure by esophagoscopy, found no correlation among the size of esophageal varices, the pressure in the varices, and the occurrence of hemorrhage. [illegible]man ([illegible]0) could find no correlation between the portal pressure and the size and extent of varices as judged on the basis of roentgenologic findings and the occurrence of hemorrhage. The risk of bleeding from esophageal varices cannot be estimated from

the portal pressure or from the size and extent of the varices Our experience suggests that a bout of acute gastritis or upper respiratory infection may have been a precipitating factor in the hemorrhage

Symptoms

The clinical manifestations are summarized in Plate XIII The signs and symptoms of the underlying liver disease should be differentiated from those of portal hypertension *per se*

Esophageal varices produce symptoms only when they bleed They do not produce obstruction Chronic or massive variceal bleeding results in anemia fatigue, and dyspnea Douglass and Snell (16), in studying 444 patients with Laennec's cirrhosis found hematemesis or melena in 32 per cent In 10 per cent massive esophagogastric hemorrhage was the initial symptom

Diagnosis

History

Gastrointestinal hemorrhage is the most important single clinical feature in diagnosis Other symptoms are usually due to the underlying liver disease (Plate XIII) The hematemesis is usually greater than that of duodenal ulcer The patient may mention an unexplained gain in weight or increase in girth He may or may not have noted enlargement of the liver or spleen or both Icterus is sometimes the presenting complaint Occasionally a history of hepatitis, homologous serum jaundice and alcoholism may be elicited

Examination

On examination jaundice cutaneous arterial angiomata and dilated abdominal veins may be observed (Plate XIII) Hepatomegaly splenomegaly and ascites may be found Edema is usually

not present although the combination of hypoalbuminemia and salt retention which is associated with the intra-abdominal pressure from ascites may result in edema of the lower extremities

X-*ray* Careful barium x-ray study using fluoroscopy and spot-film technic usually reveals multiple filling defects produced by the dilated distended veins A typical picture of extensive varices is shown in Figure 53 During early phases of portal hypertension,



FIGURE 53 X ray appearance of esophageal varices in a patient with cirrhosis of the liver who had had severe rectal bleeding with tarry stools and anemia

however, varices may not be evident at roentgenologic examination Likewise, examination during the phase of acute bleeding requires particular care and skill (Fig 54)

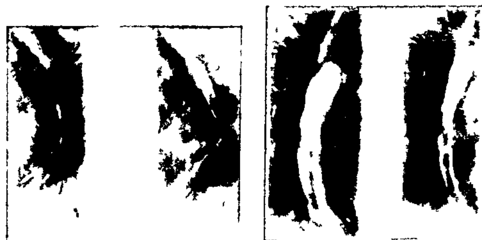


FIGURE 54 These x rays demonstrate the importance of thorough roentgenological examination This patient with hematemesis and cirrhosis, had a normal appearing barium swallow when examined from the left side (x ray at left) When the patient was examined from the right (x ray at right) the varices not visible on the previous inspection were demonstrated

Esophagoscopy is of great value in the diagnosis of esophageal varices The radiologist may miss early varices, or he may be in doubt even when they are obvious at esophagoscopy Varices seen through the esophagoscope (Plate IX, #3, Plate XIV) appear as characteristic bluish tortuous masses of veins usually involving the lower segment of the esophagus but sometimes extending its full length In the early phase however, they may not appear blue and may not be tortuous hence it is sometimes difficult to distinguish them from esophageal folds or gastric rugae in a hiatus hernia Although they may seem to obliterate the lumen of the esophagus they never cause esophageal obstruction or interfere with the passage of the esophagoscope Esophagoscopy presents no risk if carried out gently without other instrumentation We have abandoned the injection of sclerosing solutions into esophageal varices since we found the technic hazardous and the results unsatisfactory

Gastroscopy also may be of value in establishing a diagnosis of gastric varices

Laboratory tests

There are no practical laboratory tests for the diagnosis of esophageal varices The determinations utilized are liver function tests these are of diagnostic and prognostic value in both the acute and the chronic phases of esophageal variceal hemorrhage

In massive hematemasis which constitutes an emergency gastro duodenal ulceration and esophageal varices are the primary considerations in that order Unfortunately if either ulcer or varices are definitely identified one still cannot be certain that either is the source of bleeding since 10 per cent of patients with cirrhosis also have gastric or duodenal ulceration (5-) Some help may be obtained from a bromsulfalein test Zamcheck (61) found only an occasional abnormal value in bleeding peptic ulcer but noted that all patients with bleeding varices had abnormally high retentions We have found the rapid blood ammonia test described by McDermott (33) of even greater value In this test the reagents are standardized so that a blood ammonia concentration greater than 100 mcgm per 100 cc of blood produces an obvious change in color In a series of over 100 cases of massive gastrointestinal bleeding seen in the Emergency Ward of the Massachusetts General Hospital this test has not yet failed to differentiate gastro duodenal and esophageal variceal bleeding

The execution of this test is diagrammed in Figure 55 The basis of this test is the fact that the diseased liver is unable to deaminate the absorbed nitrogenous split products of gastrointestinal hemorrhage These are present as ammonia in the circulating blood stream and are recirculated through the collateral esophageal varices thus causing elevations of blood ammonia values that are detectable by means of this test

Numerous liver function tests are available but in view of the tremendous functional reserve of the liver a battery of such tests

is of more value as yielding an aggregate liver "profile" than any single test

In general we have utilized the liver-function tests as indices of the severity of liver disease rather than as diagnostic aids By the

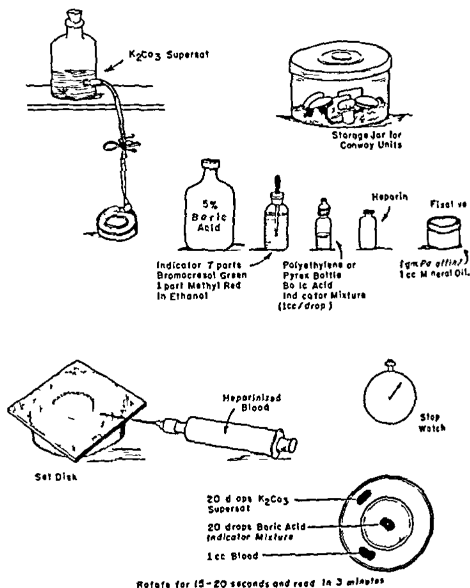


Figure 55 Rapid blood ammonia test for diagnosis of bleeding esophageal varices (McDermott W V, Jr *New England J Med* 257 1161 1957) The reagents are standardized so that if the blood ammonia level is greater than 100 mcgm per 100 cc of blood the indicator color will change from gray pink to blue

time such tests have been ordered the diagnosis of portal hypertension is usually well established. We have found these tests of value in preparing the patient for surgery and in assessing operability.

A low serum albumin (below 3.0 gm per 100 cc of serum) regularly indicates a poor prognosis; however, a high serum albumin does not guarantee against postoperative liver failure. Patients with a preoperative bromsulfalein retention of 10 per cent or less tolerate surgery well almost without exception. At this hospital (18) marked disturbances in serum bilirubin and prothrombin time were associated with a frequency of approximately 33 per cent liver failure during the postoperative course.

Liver biopsy

Whenever the diagnosis remains in doubt after thorough evaluation, liver biopsy (24) performed by aspiration with a needle may provide valuable information. This usually helps to distinguish between intra- and extrahepatic biliary obstruction; it also may pro-

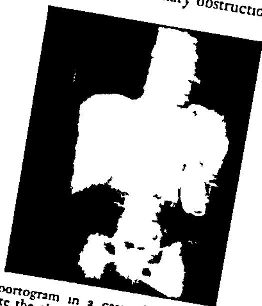


FIGURE 56. Splenoportogram in a case of splenomegaly without portal hypertension. Note the absence of collaterals.

vide a clue to the nature and the severity of the cirrhosis and may occasionally reveal a metastatic tumor

In emergencies a history of alcoholism and cirrhosis physical findings of splenomegaly and hepatomegaly and the presence of cutaneous spider angiomata or peri-umbilical collateral veins strongly suggest cirrhosis and portal hypertension Varices may be demonstrated by esophagoscopy or x ray or both

When an emergency does not exist, percutaneous splenoportography as originally described by Abeatici and Campi (1) may provide valuable information Portal pressure may be measured directly through the puncture needle and injection of a radio-opaque medium such as a 70 per cent solution of Diodrast will delineate the portal venous system and any collaterals Figures 56 and 57 demonstrate two typical splenoportograms Splenoportography with measurement of splenic pulp pressure has been advocated as an emergency diagnostic measure We consider that its risks may outweigh its advantages in such circumstances and we have utilized it only in selected cases before scheduled surgery

Treatment

Many patients with portal hypertension will probably do well under modern medical management Obviously any hospital or surgical series represents a selected group Nevertheless once hemorrhage has occurred the prognosis for life is poor The history of esophageal bleeding in a patient with portal hypertension is the chief indication for surgical treatment Since 76 per cent of patients may die at the time of the first hemorrhage (58) it is obvious that only those whose liver function was adequate to protect them against the effects of bleeding could come to operation

Emergency management

A patient with upper gastrointestinal bleeding should be hospitalized immediately and his condition stabilized by blood transfu

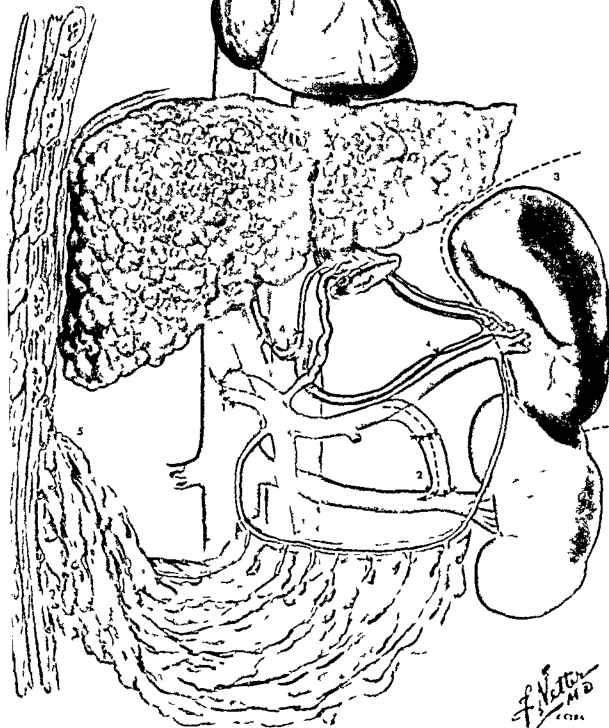


Plate XX Surgery for portal hypertension (Courtesy The CIBA Collection of Medical Illustrations, Volume 3, Part III)





PLATE XXI. Carcinoma of esophagus. (Courtesy Fletcher D. Woodward, M.D. Clinical Symposia 8:3, 1956.)



FIGURE 57 Splenoportogram in a case of cirrhosis with portal hypertension Note the extensive collateral circulation

sions Fresh transfusions should be utilized whenever possible to help to control the bleeding defects frequently associated with "hypersplenism An emergency barium swallow with fluoroscopy and spot films should be carried out in an attempt to visualize esophageal varices If these are demonstrated in the absence of

other causes of bleeding, it may be assumed that they are the source of hemorrhage Temporary control of hemorrhage is obtained with an intragastric balloon which produces cardioesophageal tamponade The tube provides both a therapeutic and a diagnostic maneuver With bleeding controlled, the patient s general condition can be more carefully evaluated, and necessary therapeutic measures such as transfusion, digitalization etc may be instituted promptly During this period further diagnostic meas ures and liver-chemistry studies may be carried out

Transthoracic transesophageal varix ligation

Because of the frequency of recurrent bleeding after removal of the tube, we proceed with transthoracic transesophageal varix liga tion (31) as soon as the patient s condition will permit surgery The patient is taken to the operating room with the balloon tube in place and the lower esophagus is exposed through a left tho racotomy incision A vertical incision is made through the fundus of the stomach and lower esophagus and the varices exposed These are usually found as three columns of dilated and tortuous veins The varices are sutured with 2–0 atraumatic chromic cat gut by a continuous over and over suture in two layers (Fig 58) Care is taken to include varices extending downward into the gastric submucosa Closure of the esophagus is carried out in the standard fashion with fine interrupted silk sutures

Preparation for definitive surgery

Because of the frequency of post suture bleeding we do not regard varix ligation as a definitive procedure The patient is al lowed a two to four week period of convalescence and prepara tions are made for some kind of shunting procedure Although it is possible to proceed immediately to an emergency shunting pro cedure once hemorrhage has been controlled by the balloon tube our experience with such a routine has been unsatisfactory We

are inclined to believe that portacaval shunts should not be carried out as emergency life-saving measures but as carefully planned procedures at a time of election

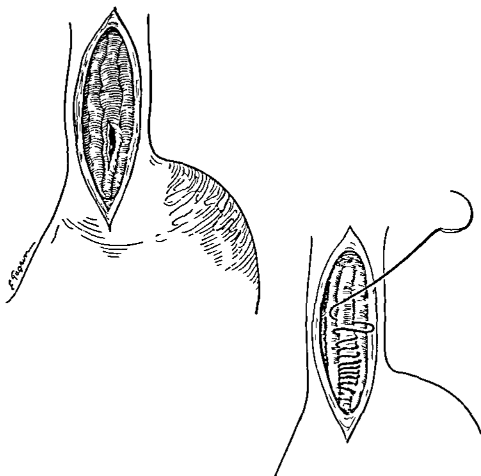


FIGURE 58 Transthoracic esophagotomy with exposure of acutely bleeding varices Suture with a running stitch of atraumatic chromic catgut

During the patient's convalescence from transesophageal suture he is provided with a high caloric intake and liver function tests are re-evaluated An attempt is made to bring his serum albumin to at least 3 grams per cent and any ascites present is treated medically Any such finding as a refractory serum albumin

level, a 3- or 4-plus cephalin flocculation, a prolonged prothrombin time after Vitamin K treatment, an elevated serum bilirubin, or a bromsulfalein retention of more than 30 per cent is a relative contraindication to shunt surgery Such a decision however, must be based on the combined opinions of internist and surgeon, with not only chemical findings but the patient s over all clinical condition being taken into account If there is any evidence of active liver disease or if needle biopsy reveals active necrosis, surgery must be postponed

Definitive surgery

A great variety of procedures have been utilized in an attempt to control varix bleeding secondary to portal hypertension Some of the earlier maneuvers attacked the source of bleeding without any attempt at influencing the primary portal hypertension

(1) Local compression — tamponade (50 59)
(2) *Injection of sclerosing solution via esophagoscope* (14 36)
(3) Intraesophageal ligation (9, 15)
(4) Partial esophagogastrostomy (43)
(5) Ligation of peri esophageal veins (23)
(6) Gastric transection (56)
(7) Mediastinal packing (54)
(8) Esophagectomy (13)

Other efforts (Plate XV) have been directed at correcting the portal hypertension

(1) Splenectomy
(2) Splenic artery ligation
(3) Hepatic artery ligation (4 45)
(4) Omentopexy (17 55)
(5) Portacaval shunts (8 48)

Currently some form of portacaval shunt is considered the optimum definitive surgical treatment The establishment of such *shunts to reduce portal hypertension* had been carried out at the turn of the century by Vidal (57), de Martel (3) and Rosen

stein (46) Their results were discouraging Whipple (60) and Blakemore and Lord (8) successfully performed such shunts and to them must go the pioneer credit in this field Their technic utilized a vitallium tube for the anastomosis this however was soon abandoned because of the high incidence of thrombosis and an everting suture technic was substituted (5 7) Satinsky (49) described the thoracoabdominal approach and Linton and Ellis (29) advocated the preservation of the left kidney by making the splenorenal anastomosis end to side Linton *et al* (30) were also the first to utilize hypotensive spinal anesthesia in shunting procedures

In general we have performed either an end to side splenorenal shunt with preservation of the kidney or an end to side anastomosis between the stump of the portal vein and the vena cava The technics for these procedures are diagrammed in Figures 59 and 60 The decision whether to perform a splenorenal or a porta caval shunt is sometimes dictated by local anatomic considerations (*e g* , portal vein thrombosis) but more frequently by the surgeon s

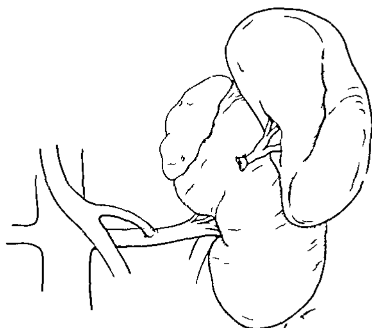


FIGURE 59 The completed splenorenal shunt

experience Preoperative splenoportography is frequently invalu-able in defining the former situations In general if the spleen is enlarged in patients with cirrhosis, one will find a large splenic vein which can be used to construct a satisfactory splenorenal shunt (27) Either procedure will usually provide a satisfactory result with control of hemorrhage if the shunt remains patent The portacaval shunt has the mechanical advantage of being larger and less likely to become angulated and technically it is usually easier to accomplish It has the theoretical disadvantage of being too large a shunt and thus entailing the complications of hepatic failure (10 26, 38)

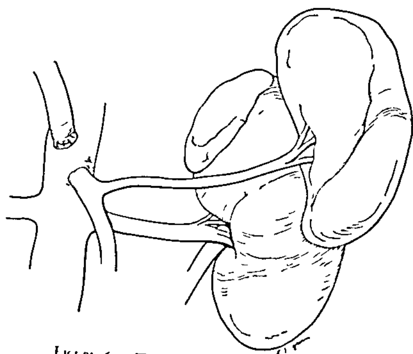


FIGURE 60 The completed portacaval shunt

Ekman (20) visualized shunts radiographically In patients who had undergone portacaval anastomosis the shunt was found to be patent in every case the portal pressure had been reduced and there was no further bleeding Splenorenal shunts resulted in control of hemorrhage in 60 per cent of his cases In 7 patients with a patent splenorenal anastomosis the size of the varices decreased in

5. This decrease was not as pronounced as in the patients with portacaval shunts. The difference in results in his series was due to the occlusion of the anastomosis in the splenorenal group. It is fair to state that a patent shunt will provide control of hemorrhage and regression of varices. There are exceptions to this, however, for we have seen more than one patient exsanguinate who was found to have a technically perfect shunt at autopsy. One patient bled to death with both a patent splenorenal and a portacaval shunt, the latter having been performed on the assumption that recurrent bleeding was due to thrombosis of the former.

The combination of fresh blood transfusion at the time of surgery and hypotensive spinal anesthesia (21) has nearly eliminated bleeding during surgery.

Recent authors (20, 58, 40, 41) have suggested that the presence of esophageal varices *per se* with previous hemorrhage constitutes a proper indication for surgical portal decompression. This has been questioned by Nachlas *et al.* (37). As with the evaluation of shunt surgery *per se*, more experience will be necessary before sound conclusions can be made concerning the role of prophylactic shunt surgery.

Complications

One of the commonest complications during the acute phase of hemorrhage is aspiration pneumonitis due to hematemesis. Proper precautions should be taken against this and a cuffed endotracheal tube utilized for all surgical procedures.

The development of hepatic coma owing to failure of the liver to detoxify split products of protein metabolism is an ominous and frequently fatal complication. It is usually a sign of recurrent or continued gastrointestinal bleeding and it must be recognized early and confirmed by blood ammonia studies so that proper treatment may be instituted. Therapeutic measures consist of enemas, gastric lavage through a balloon tube, oral administration of antibiotics (preferably neomycin) and intravenous injection of

glutamic acid or arginine The reader is referred to the papers of McDermott (34) for a thorough review of this problem

Prognosis

Esophageal varices associated with portal hypertension can cause death by exsanguinating hemorrhage At the Massachusetts General Hospital 45 per cent of the deaths in patients with cirrhosis and 71 per cent of the deaths in those with Banti's syndrome (subhepatic obstruction) were caused by bleeding esophageal varices (31) Conversely, it has been estimated that 50 to 70 per cent of patients with portal hypertension will bleed to death within one or two years after their first hemorrhage (6, 28, 44 51)

In comparing the mortality of repeated hemorrhages in patients who had varices with and without cirrhosis Merendino and Volweiler (35) *found the mortality rate five times greater among* those with cirrhosis Seventeen per cent of patients died after their first hemorrhage

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Benign Tumors and Cysts

18

Any tumor which does not metastasize is usually referred to as a benign tumor Benign tumors include papilloma adenoma angioma myoma fibroma lipoma myxoma edematous polyps and cystic growths

Incidence

Benign tumors of the esophagus are rare A survey of the literature since 1933 reveals reports of only 250 cases of benign tumors and cysts of the esophagus (6) By far the commonest neoplasm of the esophagus is leiomyoma Lewis and Maxfield (10) were able to collect 155 cases in which there were a total of 19 tumors One hundred and eight were found at post mortem and 47 at operation Probably many cases have never been recorded and some in the literature have undoubtedly been overlooked The rarity of benign neoplasm of the esophagus has been further emphasized by these authors who state that deaths from esophageal malignancy in the United States alone during 1949 were 3933 which is 6 times all the reported cases of myoma in all

the years since 1717. According to their figures, 46 per cent occurred in the lower third of the esophagus, 35 per cent in the middle third, 9 per cent in the upper third, 8 per cent in the esophagogastric region, and 2 per cent in the cervical segment. Moersch and Harrington (13) reported 44 cases in 7459 necropsies (0.59 per cent). In 6001 post mortem examinations at the University of Chicago 11 benign esophageal tumors (0.18 per cent) were found by Schafer and Kittle (18). According to Adams and Hoover (1) the number of cases discovered and recorded during the last 11 years is over one-half as large as the number of cases recorded in the preceding 220 years. This increase is attributed to improved x-ray technic and esophagoscopy.

Etiology

The etiology of benign tumors of the esophagus is not known. It is possible that some of them arise from embryonic rests. They originate from smooth muscle, connective tissue, and nerve sheath, and in cysts resulting from bronchial or esophageal cell rests.

Pathology

Benign tumors are divided into two main groups — mucosal and extramucosal. The former type arises from the mucosa or submucosa generally in the upper portion of the esophagus and grows into the lumen, where it tends to become pedunculated. The latter type arises from the extramucosal layers and grows within the wall. Generally these develop in the lower half of the esophagus. Although they are not always encapsulated, they are not invasive.

Benign tumors vary greatly in size. Sometimes they are very small and produce no symptoms, but quite often they are large enough to cause marked obstruction of the esophagus. In 1953 Kenney (9) reported the case of a very large esophageal leiomy

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18

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Incidence

Benign tumors of the esophagus are rare. A survey of the literature since 1933 reveals reports of only [illegible]50 cases of benign tumors and cysts of the esophagus (6). By far the commonest neoplasm of the esophagus is leiomyoma. Lewis and Mayfield (10) were able to collect 155 cases in which there were a total of 19 tumors. One hundred and eight were found at post mortem and 47 at operation. Probably many cases have never been recorded and some in the literature have undoubtedly been overlooked. The rarity of benign neoplasm of the esophagus has been further emphasized by these authors who state that "deaths from esophageal malignancy in the United States alone during 1949 were 3933, which is 6 times all the reported cases of myoma in all

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oma weighing 14.0 gm. It was thought to be the largest esophageal tumor ever resected surgically. In 1955 Lodmell (11) reported the case of a giant benign fibromatous polyp. The gross specimen was 25 cm long and a pedicle was present at the upper end. The tumor gradually enlarged from approximately 4 cm at the upper part to 6 cm in diameter distally.

DiMatteo (6) classifies the origin of benign tumors as follows: (1) mucosal—adenomas, epithelial cysts and hemangiomas, (2) intramural—leiomyomas, lipomas, and fibromas, and (3) paraesophageal—chronic granulomas and embryonic rests. Cornell, Blumberg and Sarot (5) classify cysts of the esophagus as reduplication cysts of the alimentary tract, ciliated epithelial cysts, cysts arising from islands of gastric mucosa, cystic or follicular esophagitis and retention cysts of ducts and mucous glands. In the series of leiomyoma cases collected by Lewis and Maxfield (10) multiple tumors were encountered in 9 per cent of the autopsy cases and 4 per cent of the operative cases, the over-all occurrence being 7.7 per cent. Nine of the 12 cases with multiple lesions had two growths, 1 had four, and 1 operative and 1 autopsy case each had fourteen. In 5 autopsy cases the myoma was found in an esophageal diverticulum.

The question whether *malignant sarcoma* can arise from preexisting benign myoma has often been raised. On the basis of 9 cases of leiomyosarcoma reported in the literature (7, 12) there was no proof that malignant transformation occurred, but the possibility could not be excluded. Concrete evidence that sarcomatous degeneration can take place is furnished by the finding of areas of sarcoma in otherwise typical clinical and histological myomas in the fifth autopsy case of Bezza (1932) (2) and the operative case of Biasini (1949) (3).

Symptoms

Small tumors usually produce no symptoms whatsoever; large ones produce obstruction with dysphagia and regurgitation. Dys-

phagia is the most common complaint, though not always the chief symptom Pain or discomfort in the retrosternal retroxiphoid or epigastric region is the second most common symptom and it often accompanies the dysphagia Vascular or ulcerating tumors may bleed with resultant melena or hematemesis If there is frequent regurgitation cough may be a prominent symptom because of overflow into the larynx Pedunculated growths at the upper end of the esophagus may be vomited into the buccal cavity or even extruded beyond the lips Such pedunculated tumors may cause irritation of the larynx with cough Rarely such a pedunculated tumor may cause death by strangulation when the tumor has been aspirated into the trachea Digestive complaints such as belching nausea, and anorexia may be noted Weight loss sometimes occurs owing to restricted dietary intake

Diagnosis

Radiology

The diagnosis of benign tumor of the esophagus cannot be made by means of history physical examination or laboratory studies X-ray examination is of fundamental importance It usually reveals a smooth filling defect at a certain level of the esophagus with normal mucosa often overlying the tumor mass Under such conditions the radiologist should suspect an intramural extramucosal benign type of tumor (Figs 61 6 see Plate IX #4) A smooth polypoid shadow with a pedicle is characteristic of a pedunculated benign tumor Sometimes the cystic nature of benign esophageal tumor can be suspected on the basis of changes in the shape of the mass under fluoroscopic examination

Esophagoscopy

Although an exact histologic diagnosis can be made only by esophagoscopy and biopsy frequently the surgeon is satisfied with

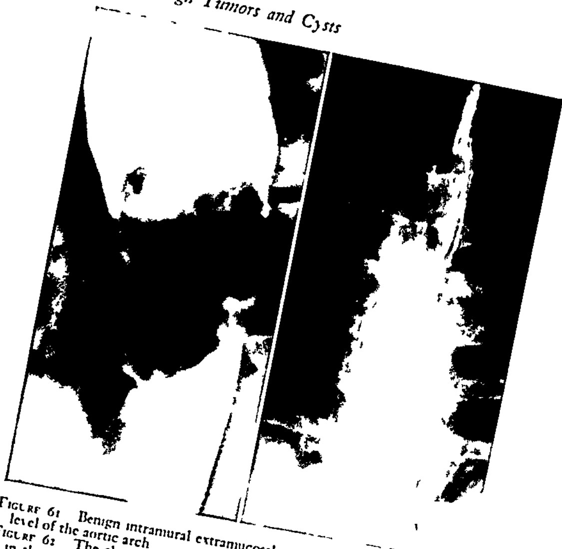


FIGURE 61 Benign intramural extramucosal tumor of the esophagus at the level of the aortic arch

FIGURE 62 The above x ray was reported as showing an obstructing mass in the mid third of the esophagus. In spite of some peculiarities cancer was considered the most likely diagnosis The radiologist considered it possible that the esophagus was involved secondarily from the outside and advised bronchoscopy to check the left main bronchus. Esophagoscopy with biopsy proved leiomyoma

the x ray diagnosis of benign esophageal tumor and considers that esophagoscopy is unnecessary and that surgical removal is essential hence in many cases esophagoscopy may be omitted Furthermore esophagoscopic biopsy of a benign tumor is hazardous and usually is contraindicated.

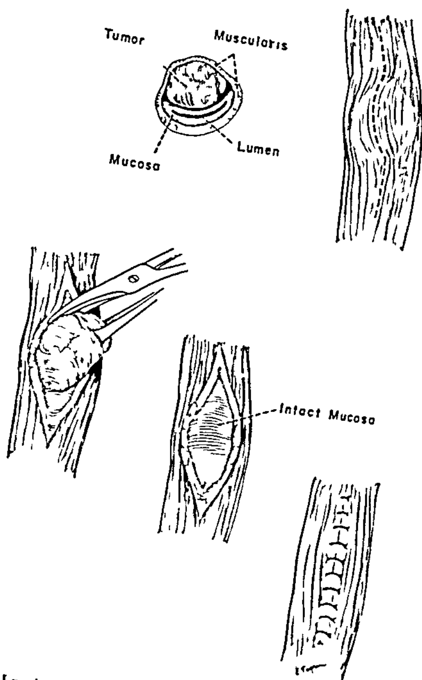
If however the diagnosis of benign tumor is not established by x-ray examination esophagoscopy should be carried out Smooth pedunculated tumors may readily be recognized by their gross appearance A rounded submucosal mass has a very different appearance at esophagoscopy from an irregular nodular carcinomatous mass In leiomyoma the mucosa usually appears normal and a tumor mass is seen to be obstructing the lumen A small intramural tumor sometimes cannot be observed by the esophagoscopist Usually however, it can be seen in the wall of the esophagus or may be described as extra esophageal The lumen may be narrowed by the leiomyoma but there is no obstruction to the passage of the esophagoscope since there is no definite stenosis The elasticity of the uninvolved esophageal wall is maintained, furthermore, the tumor is usually found to be freely movable

TREATMENT

Because of the danger of hemorrhage or perforation of the esophageal wall only a few benign tumors of the esophagus can be removed by esophagoscopy Such tumors are the nonvascular pedunculated tumors the pedicles of which may be severed with cutting forceps Although such removal has been reported we believe that surgery of the esophagus is now so safe that one should have no hesitation in referring a good risk patient to a thoracic surgeon In this way the danger of hemorrhage perforation, or possible malignant degeneration is avoided

Sauerbruch (17) is generally credited with the first successful removal of an esophageal cyst in 1937 although Ohsawa (14) and Krauss (8) both reported successful excisions in 1933

The majority of benign tumors encountered by the surgeon are leiomyomas usually found in the lower third of the esophagus (15) They may be multiple Cysts of esophageal and bronchial origin are the second most common group



...URE 63 Enucleation of benign intramural esophageal tumor Care is ...aken not to incise the mucosa

As previously mentioned esophagoscopy is rarely indicated in such cases and if it is performed the endoscopist should not take a biopsy specimen from a submucosal tumor, since the resultant fibrosis makes subsequent enucleation difficult and even hazardous

Hemangiomas of the esophagus are not amenable to surgical treatment There are no reported successful resections (4)

Because of their possible malignancy and tendency to grow benign tumors in the good-risk patient should be resected when diagnosed, whether or not symptoms are present

The *treatment of choice is simple enucleation without penetra*tion of the mucosa followed by reconstruction of the esophageal wall (Fig 63)

If the mucosa is torn or broken into it should be carefully reconstructed with interrupted stitches of fine silk with knots tied on the inside Care must be taken not to narrow the lumen

In most cases these tumors may be approached through either side of the chest If the tumor is large or is so situated that simple enucleation may not be adequate a left thoracotomy should be *utilized so that an esophageal resection with esophagogastrostomy* (Chapter 19) can be performed This however is rarely necessary

Prognosis

The prognosis of benign tumors of the esophagus when properly treated is excellent

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Carcinoma

19

Carcinomas of the esophagus (Plate XVI) are malignant tumors usually arising from squamous epithelium

Incidence

Carcinoma is one of the most important diseases of the esophagus It is next in frequency to esophagitis and achalasia It has been stated by Adams ([illegible]) that it ranks fourth in incidence of all malignant tumors in men over [illegible]0 years of age Although death has occurred from cancer of the esophagus in a child only 1 year old (19) it is usually a disease of middle and late life It is much more common in men than in women in the approximate ratio of 4 to 1 however in the post cricoid area it occurs very much more commonly — in fact almost exclusively — in women ([illegible]) It accounts for 5 to 7 per cent of all carcinoma in males (38) According to Shedd Crowley, and Lindskog (38) the age distribution in 180 patients was as follows under 40 years 1 patient between 41 and 50 years 11 patients 51 to 60 years 57 patients 61 to 70 years 69 patients 71 to 80 years 40 patients over 80 years [illegible] patients There were [illegible] females (1[illegible] per cent) and 158 males (8[illegible]

per cent) There were 174 white and 6 Negro patients Franklin and Shipman (13) studied 59 patients with carcinoma of the esophagus and found the age range to be 22 to 81 years with 65 per cent in the seventh and eighth decades (average age 63)

In this chapter we are discussing primary carcinoma of the esophagus but it is interesting to note that carcinoma of the fundus of the stomach often infiltrates the lower portion of the esophagus In Humphreys and Moore s series (20) there were 162 cases of primary carcinoma of the esophagus as compared with 47 cases arising from the stomach and invading the lower segment of esophagus Gibbon *et al* (16) also found primary carcinoma of the esophagus to be about three times as common as carcinoma arising in the stomach and invading the esophagus (67 to 22)

Etiology

The etiology of carcinoma is unknown Chronic irritation may be a factor in some cases One of us (4) reported 2 cases of carcinoma of the esophagus developing in benign stricture (Figs 64 65) these cases comprised one of lye stricture and one of congenital stenosis Including these two we now have a total of 9 cases of carcinoma which developed in pre-existing benign lesions (23) as follows web of upper esophagus 2 web of lower esophagus later becoming a benign inflammatory stenosis 1 esophageal ulcer with benign peptic stenosis (19 years duration) 1 achalasia 2 lye stricture 2 congenital stenosis (60 years duration) 1 These benign lesions were all of many years duration and they tend to support the theory of chronic irritation as an etiological factor

Other writers have made similar observations For example Kay and Cross (24) found that carcinoma of the esophagus developed in 10 patients with chronic esophagitis 7 of whom had an associated hiatus hernia 2 a pulsion diverticulum of the lower esophagus and 1 achalasia of the esophagus The 7 patients were from a group of 200 with hiatus hernia The 2 patients with pulsion diverticulum of the lower esophagus were from a group of

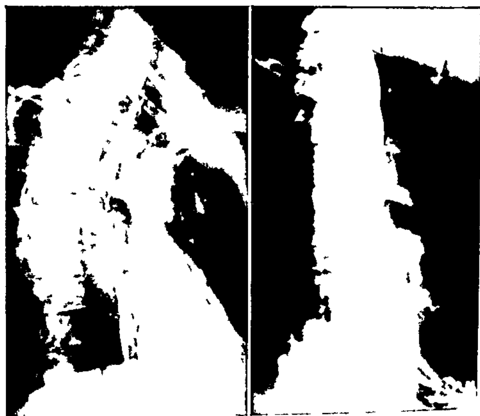


FIGURE 64 Multiple strictures of the esophagus. The upper stricture was a carcinoma which had developed 29 years after the ingestion of lye (Courtesy of the *New England Journal of Medicine*)

FIGURE 65 X ray appearance of carcinoma of the lower esophagus which developed in a man 57 years old who had had symptoms of esophageal obstruction all his life probably due to congenitally short esophagus and benign stricture (Courtesy of the *New England Journal of Medicine*)

12 with this condition The one patient with achalasia and carcinoma of the esophagus belonged to a group of 75 patients with achalasia Wiley (49) states that the discovery of a hiatus hernia in association with a lower esophageal or upper gastric carcinoma is not uncommon A hiatus hernia was found to coexist with esophageal cancer in 10 of 100 patients observed by him between 1948 and 1954 Bullock and Snyder (6) reported carcinoma *in situ* occurring in pharyngeal diverticulum Mosbech and Videbaek (31) made a special study of the etiology of esophageal carcinoma in 101 patients including 83 men and 18 women Hereditary factors

apparently played no part Abuse of alcohol seemed to be important at least in men, as 65 per cent of patients had been alcohol addicts and 21 per cent had been occupationally exposed to alcohol No correlation was found between syphilis and carcinoma of the esophagus (such as exists between syphilis and carcinoma of the tongue)

Wurnig (52) made a similar study of conditions predisposing to carcinoma of the esophagus he found 12 cases among 198 in which it could be assumed that chronic benign disturbances had caused the development of cancer These cases included 1 case of traction diverticulum and 7 cases originating in lye corrosion, other cases included 1 of cardiospasm with accompanying chronic esophagitis and 3 were cases of shortened esophagus It is conceivable that tuberculosis of the esophagus — which is extraordinarily rare — might play a part in the development of carcinoma as is suggested by a case of carcinosarcoma of the esophagus reported by Thompson (45) in which several early tubercles were found about the periphery of the tumor

Pathology

Carcinoma of the esophagus may develop at any level but is commonest in the middle third and only slightly less common in the lower third Carcinoma of the upper third is much less frequent In the upper and middle thirds it is likely to be of the squamous cell type whereas in the lower third it may be either squamous cell or adenocarcinoma this is probably related to the common occurrence of gastric mucosa in the lower end of the esophagus Carcinoma of the fundus of the stomach frequently invades and infiltrates the lower segment of the esophagus According to Smithers (39) adenocarcinoma of the esophagus has been said to arise (1) from ectopic islets of gastric mucosa or in esophageal glands (2) in esophageal mucous membrane of which a section has failed to undergo squamous transformation before birth (3) in mucosa which has undergone some glandular meta

plasia after attrition or long standing infection (4) in minor extensions or folds of gastric mucosa lining the hiatal canal, (5) in the thorax distal to the cardia in a tube-like extension of the stomach in a patient with a congenitally shortened esophagus, and (6) from primary tumors of the stomach usually in the esophageal submucous lymphatics. Smithers summarizes his own experience however, as follows "Twenty-three adenocarcinomas of the esophagus, not associated with hiatus hernia are reported in addition to 7 with that association previously published and 1 further case recorded here. It is suggested that such tumors usually arise in lower esophageal mucous membrane which has failed to undergo squamous transformation before birth."

Any *carcinomatous growth in the esophagus may be proliferating* ulcerating or infiltrating. Many carcinomas seem to be a combination of all three. The *infiltrating type* are likely to be annular and stenosing whereas the proliferating type may be soft and polypoid and may cause comparatively little obstruction. On the other hand large firm cauliflower like masses always produce some obstruction. Most carcinomas of the esophagus are highly malignant invade neighboring structures and metastasize early to regional lymph glands submucosal lymphatics and distant parts of the body notably the liver. Of particular interest to the endoscopist is direct extension to the trachea or left main bronchus. Such invasion may occur when *the lesion is in the middle* or upper third of the esophagus rendering it probably incurable. Such a *situation may* be *predicted* by means of bronchoscopy (Plate IX, #8).

Symptoms

Unfortunately in the early stages carcinoma of the esophagus produces no symptoms whatsoever. Even in the late stages [illegible]0 per cent of cases exhibit a non stenosing variety *of lesion* (10). The characteristic symptoms of the stenosing type are dysphagia pain *regurgitation and vomiting*. Dysphagia is the earliest and

most frequent complaint in this type of tumor Loss of weight is a common symptom but not an early one More than 50 per cent of patients have no pain If pain is present at all it may occur only during swallowing It may be sharp dull or boring and may be either continuous or intermittent it is likely to be substernal or epigastric Pain in the back is an occasional symptom this has bad prognostic significance since it often means posterior local invasion Aspiration of regurgitated food or secretions may lead to cough Hoarseness is present in a few cases owing to pressure on the recurrent laryngeal nerve Unusual manifestations of carcinoma of the esophagus have been studied recently by Wohl Pastor and Karr (50) who also emphasize the fact that 20 per cent of esophageal carcinomas are non-obstructive when first seen in these cases dysphagia is absent These workers report 7 cases with the presenting complaints of hoarseness, cough lung abscess pneumonia hematemesis enlargement of the thyroid gland and supraclavicular mass In many instances the symptoms were due to distant metastases Gross bleeding in the form of either hematemesis or melena is unusual but occult blood may be found in the stools Although the disease is a progressive one the dysphagia may be intermittent in character, this is related not only to the different types of food eaten at various times but also to the amount of edema of the tumor and to the amount of food previously caught in the tumor mass A variable amount of spasm is present in the esophagus just above the tumor Unfortunately the patient may not have much difficulty in swallowing solid foods and he may postpone consulting a physician until he can take nothing but very thin liquids

Complications

If they occur at all complications are usually seen late in the disease Perforation into the mediastinum may occur with localized abscess formation or mediastinitis Perforation into the tracheobronchial tree is common late in the disease and results in esoph-

ageotracheal or esophagobronchial fistula. On rare occasions the growth may penetrate the aorta, with resultant fatal hemorrhage into the esophagus.

Diagnosis

Carcinoma of the esophagus should be suspected in every patient *presenting himself with a history of dysphagia.* Even the slightest amount of intermittent dysphagia must not be disregarded if the diagnosis is to be made early. Early diagnosis is, of course, the *principal factor in successful treatment.* *Other symptoms* mentioned above, do not help much in early diagnosis. Physical examination in the early stages is entirely negative.

X-ray examination

Careful x-ray examination is of the utmost importance; in a typical case it reveals an irregular filling defect in the esophagus (Fig. 66). *Sometimes, however, x-ray examination is equivocal, revealing* only an annular, smooth narrowing which might be consistent with benign stricture, spasm or carcinoma (Fig. 67). The expert radiologist *seldom misses an obvious esophageal filling defect.* Radiologists of less experience may find x-ray examination of the esophagus quite difficult to interpret; even the most experienced are sometimes baffled by their findings and therefore urge esophagoscopy. Occasionally very good radiologists may confuse esophageal varices with carcinoma. Among 1000 cases *of cancer of the esophagus* Resano (36) observed 15 cases that had not been discovered at radiologic exploration. A satisfactory roentgenographic study of esophageal *cancer should give detailed information* not only about intraluminal involvement and the length of the lesion but also about its lateral and para-esophageal extension (18). In a study of 187 cases of primary epidermoid carcinoma Parker (35) found that 6 patients who had had negative radiographic studies of the esophagus were proved *at biopsy to have carcinoma.* This author stated that since his roent

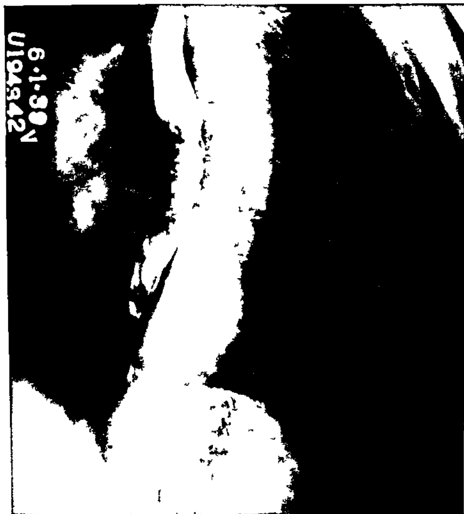


FIGURE 66 Typical irregular filling defect seen in carcinoma of the esophagus (Courtesy of the *Annals of Otology, Rhinology and Laryngology*)

genologic colleagues were highly trained and cooperative workers he was much impressed by the great need of single or multiple esophagoscopic examinations in many patients who have suggestive symptoms

Taylor (44) has reported a paraganglioma that simulated carcinoma of the esophagus in every respect In his case there was a nine year history of dysphagia due to compression of the cervical

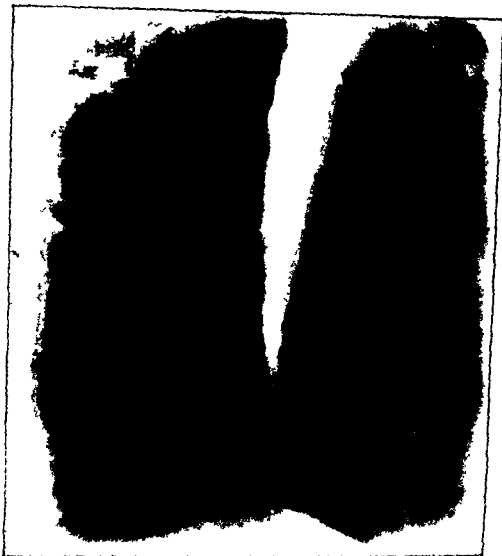


FIGURE 68 X-ray appearance of the esophagus reported as showing benign stricture Esophagoscopy done at another hospital had revealed only a smooth conical narrowing thought to be due to benign stricture No biopsy specimen was taken At a second esophagoscopy at this hospital a biopsy specimen was obtained which revealed submucosal carcinoma The carcinoma was proved to have originated in the stomach and involve the latter very extensively, in fact peritoneoscopy revealed carcinomatosis.

suspicion of such esophageal infiltration arises, esophagoscopic biopsies may yield positive evidence of adenocarcinoma

Cytology

Expert cytologists can often make a diagnosis of carcinoma of the esophagus on the basis of carefully taken esophageal washings In 40 cases reported by Klayman (25) there were no false positive diagnoses In his experience 19 of 20 cases of esophageal neoplasm were detected by exfoliative cytologic study In many clinics however there have been some false-positive reports the confusion arising especially in cases of inflammatory stenosis In this hospital we know of two such false positive reports however, we know of one important case of carcinoma developing in achalasia in which only cytologic study established a positive diagnosis (See case reports Chapter 4) It should be pointed out that in achalasia associated with megaesophagus x-ray examination and esophagoscopy may be difficult owing to retention of food in a dilated esophagus and that hence a small tumor can be missed by both methods this fact indicates the value of cytology in such a situation Confidence can be placed in the cytologic reports only in proportion to the experience of the cytologist

Treatment

We agree with Buschke (8) that both surgical and radiological methods of treatment for esophageal carcinoma are still in need of improvement Buschke found that although surgical and radiological technical facilities have improved during the last decade the five year cure of patients with esophageal carcinoma is no more than 2 or 3 per cent We are personally aware of two five year cures following x ray therapy in this hospital

X-ray therapy

When the tumor is considered definitely inoperable because of marked local fixation, invasion of vital structures or other reasons or when it has been proved *inoperable by exploratory thoracotomy* palliative treatment is certainly worth while This usually consists of x-ray treatment sometimes combined with bouginage which may occasionally permit swallowing and ameliorate dysphagia Patients in this group who are otherwise in good health with expectation of a reasonably long survival, may be candidates for feeding gastrostomy to tide them over the period of therapy during which esophageal occlusion may become temporarily complete

In irresectable low-lying lesions we have occasionally obtained excellent palliation with a side-to-side esophagogastrostomy — mobilizing the stomach above the tumor through the left side of the chest (Fig 69)

Our experiences with the Souttar tube (40) have been almost uniformly unsatisfactory We have had no experience with the plastic prosthetic devices described by *Berman (5)* and Mackler and Mayer (28)

Occasionally the primary tumor disappears entirely under high voltage radiotherapy and the patient is apparently cured — only to die later of distant metastases Permanent cures have been reported Watson and Brown (48) state that carcinoma of the esophagus can be eradicated by properly planned beam directed deep x ray therapy They reported 4 cases with clinical histories and post mortem findings in 3 of which death was due to other causes and at autopsy no residual esophageal carcinoma and no metastatic cancer was found On the other hand Krebs *et al* (26) in a discussion of the rotation x ray treatment of cancer of the esophagus state that the curative effects were slight — that in many cases the local tumor disappeared after x ray therapy but the patients later died from metastases However palliative effects such as larger food intake relief from dysphagia and improvement in

general condition were noted in more than 70 per cent of the cases

Scharer (37) at the radiotherapeutic clinic of the University of Zurich employed the rotating method for esophageal carcinoma

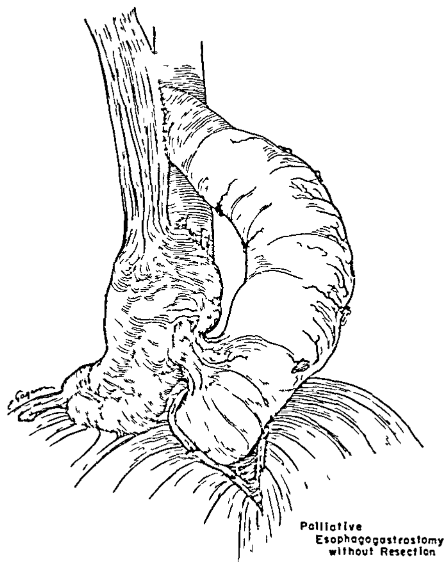


FIGURE 69 With some low lying irresectable lesions side-to-side esophagogastrostomy without resection may provide palliation and permit swallowing

unless the tumor was in an unfavorable eccentric position either at the jugular level or near the cardia Five-year cures were obtained in - per cent of the cases and three-year cures in 4 per cent Great improvement was obtained in 17 per cent of the patients Local cure was obtained *in 4 5 per cent, but these died from metastases* Fifty-seven per cent of the patients either were not helped *or experienced* exacerbation during treatment The results of rotating irradiation were only slightly better than those obtained when irradiation was applied to several areas

Routine post resection radiation therapy as suggested by Adams *et al* (1) may have a place in the treatment of carcinoma of the esophagus, but as yet, as Dr Adams admits, "it is certainly too early to tell and perhaps we will be disappointed in the end

Surgery

We believe that surgical extirpation is the treatment of choice for most esophageal carcinomas However, owing to variations *in lymphatic drainage and to the intimate association* of the esophagus with other mediastinal structures the majority of esophageal neoplasms are incurable when first seen by the surgeon There are clinical findings which suggest that the lesion has spread so far that it is no longer curable and is probably inoperable but in patients with severe dysphagia *we do not necessarily* consider that these findings always contraindicate operation Such observations include positive supraclavicular fat pad biopsy paralysis of the vocal cord Horner's syndrome tracheal or bronchial fistula metastatic disease and endoscopic evidence of fixation If operation is undertaken in a patient *demonstrating* any of these signs the surgeon must be certain that he is at least offering palliation Unless the patient's general condition is so poor that he will not tolerate anesthesia or his life expectancy is extremely short these tumors particularly those with complete obstruction should have the benefit of surgical *exploration At the time* of operation operability can be estimated and resection should be undertaken

whenever it is technically possible The inoperability of these cases does not result from nodal involvement but from impossibility of separating the esophagus from contiguous structures such as the aorta or the pulmonary hilum This can be determined only at the time of surgery If resection is impossible some form of anastomosis without resection which will permit swallowing should be attempted, so that the patient will not starve to death or drown in his own saliva Should all surgical maneuvers be unsuccessful rotational high voltage x-ray therapy or the newer cobalt treatment coupled with bouginage may occasionally provide good palliation A gastrostomy may be performed to tide the patient over his radiation therapy In general we are opposed to gastrostomy in the treatment of carcinoma of the espohagus, it does not add to the comfort of the patient and is associated with an appreciable mortality In 50 cases of carcinoma of the esophagus treated by "palliative' gastrostomy Dickison (1-) reported that -3 died in the hospital and that the average survival time of the entire group was 1-0 days during which period their distress was if anything greater than before operation

Torek (46) was the first who successfully extirpated the esophagus for carcinoma in 1913 The operation was performed on a 69 year-old woman who survived to the age of 81 Gastrointestinal continuity was not re established the patient being left with a cervical esophagostomy and gastrostomy Twenty years later Ohsawa (34) in Japan performed successful resections with anastomosis In the United States Adams and Phemister (3) and Marshall (-9) reported the first successful esophageal resections

Lower esophagus and cardia

Carcinomas of the lower end of the esophagus and cardia of the stomach (Fig 70) are technically the easiest to remove and generally provide the best end results

We have usually utilized a left thoracotomy incision through the bed of the eighth or ninth rib If the surgeon prefers a tho-

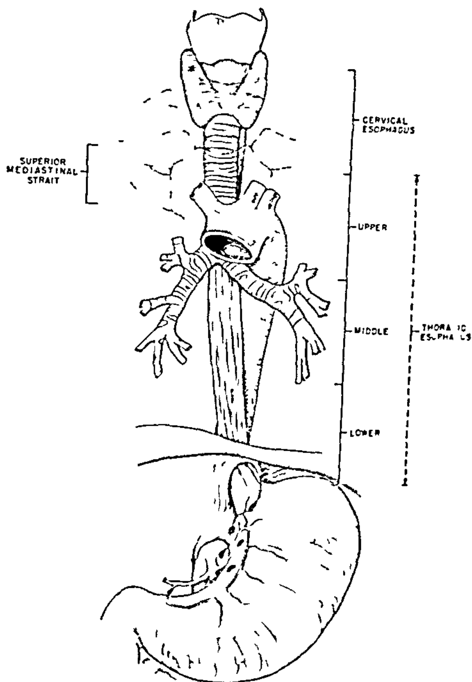


Figure – Anatomical divisions of the esophagus

racoabdominal incision may be made this may be of particular value if intraperitoneal metastases are suspected In such cases the abdominal portion of the incision is made first and exploration can be carried out before the chest is opened

After the thorax is opened the phrenic nerve is crushed the *esophagus mobilized and the vagus nerves divided well above the* proposed line of resection The diaphragm is incised radially and the abdomen explored it may be necessary to include a portion of the diaphragm with the specimen The esophagus above the tumor and the stomach below are mobilized particular care being taken to preserve the right gastric and gastroepiploic arteries The left gastric artery should be divided at its origin from the celiac axis in order to remove the large group of lymph nodes in this area

Payr clamps are then placed across the stomach to preserve as long a portion of the greater curvature as possible (Fig 71 A) The stomach is divided with a knife a dry gauze is tied over the proxi-

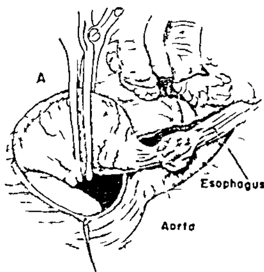
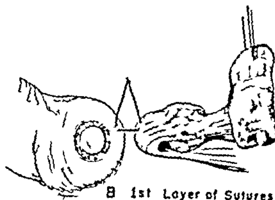


FIGURE 71 Operation for carcinoma of the lower third of the esophagus. The first drawing (A) shows the mobilization of the stomach and lower esophagus, and the use of Payr clamps. In the second drawing (B) the gastric stump has been turned in and a circular serosal incision made for the first suture layer Drawings C through F show completion of anastomosis. (See pages 24[illegible] 241)

mal end and the tumor-bearing segment is displaced from the operative field It will serve as a handle for the esophagogastric anastomosis The distal end of the stomach is closed with two running layers of catgut and a third layer of fine interrupted silk sutures.



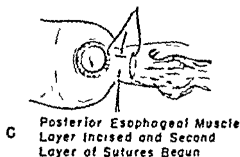
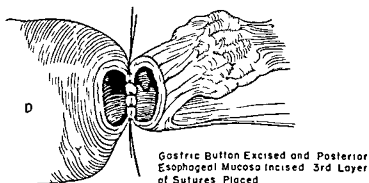
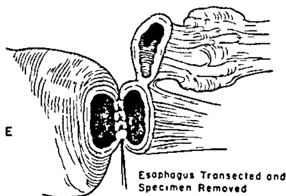


FIGURE 71 (*continued*) See legend page 239

The technic of the anastomosis is that originally described by Churchill and Sweet (10) as later modified by Sweet (43) A circular incision is made through the serous and muscular coats of the anterior wall (Fig 71B) Small bleeding vessels are stitch ligated with fine silk A three layer interrupted silk anastomosis is begun by suturing the muscularis of the esophagus to the serosa of the stomach The muscle layer of the esophagus is then incised and a second muscular layer of sutures is placed (Fig 71C) The mucosal gastric button is now excised and the posterior esophageal mucosa incised so that the third mucosal layer may be

placed (Fig 71D) These sutures are placed so that the knots will lie within the lumen when tied The distal esophagus is then completely transected the specimen is removed and the anastomosis





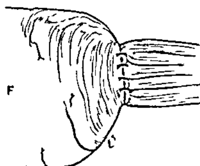


FIGURE 71 (*continued*) See legend page 239

is completed by three identical anterior layers (Fig 71D, E, F).

Care must be taken not to place the circular gastric opening used for the anastomosis too close to the gastric turn in, otherwise necrosis of the small bridged segment may result.

Tension at the anastomosis must be avoided. The stomach is fixed to the mediastinal pleura by a series of interrupted stitches and the diaphragm is reconstructed about the stomach, care being taken not to constrict it.

Mid-esophagus

Tumors of the mid esophagus may quickly become inoperable by reason of fixation to nearby structures which cannot be removed, such as the hilum of the lung or the aorta. Frequently there is lateral spread of disease toward the right mediastinal pleura. Lewis (27) and Hurley (21) have pointed out the advantages of a right-sided approach to this portion of the esophagus after preliminary mobilization of the stomach through an epigastric incision. There is no doubt that this approach has much to commend it for ease of esophageal mobilization and it is probably the technic most commonly used today. However a separate abdominal incision must be made in order to mobilize the stomach. Garlock and Sweet still favor the left thoracic approach. We have found the thoracoabdominal incisions suggested by Moore and Lawrence (Chapter 7) occasionally helpful as a compromise approach.

In general our approach to mid esophageal lesions has been made through the left side of the chest as for lower lesions. A second opening resecting the fourth rib has been invaluable in mobilizing the structures above the aortic arch and in carrying out a high anastomosis.

Although the principles of the operation in this area are identical to those for the lower esophagus several points are worthy of emphasis. Because extensive mobilization of the stomach is necessary for a supra aortic anastomosis the lateral peritoneal attachment of the duodenum should be completely divided. If this is

done it will rarely be necessary to perform concomitant pyloroplasty

Burgess *et al* (7) showed that with squamous-cell cancers there is microscopic spread in the esophageal wall up to 4 cm beyond the macroscopic edge of the tumor In order to provide an adequate margin all lesions of this area must be anastomosed above the aorta This also eliminates carrying out an anastomosis at or just below the arch where the blood supply to the esophagus may be critical (See Chapter -)

Accordingly after completion of the abdominal and lower thoracic dissection the esophagus is freed under the arch of the aorta (Fig 7-A B) Particular care must be taken at the level of the upper aortic arch for it is at this level that the thoracic duct crosses the esophagus If the duct is adherent to the tumor it is excised and the divided ends carefully ligated

The stomach is divided as close to the cardia as possible with removal of the nodes in the region of the left gastric artery The proximal segment of the esophagus is covered by rubber dam and pulled up behind the aortic arch The stomach is brought up behind the pulmonary hilus and anterior to the aorta and an esophagogastric anastomosis carried out through the higher thoracotomy incision by the previously described technic (Fig 7 C D)

Postoperative leaks from either the suture line or necrotic areas of the stomach are serious complications Carey and Clagett (9) found the commonest cause of postoperative death in their series of 131 patients was anastomotic leak Not only must the right gastric and gastroepiploic arteries be visualized and preserved but the entire gastric mobilization must be carried out gently and delicately Garlock and Klein (14) are convinced that rough handling and the use of clamps are factors in the development of localized thrombosis with subsequent gastric necrosis and leakage In 13 such leaks they found 9 due to localized gastric necrosis 3 to healing disturbances at the suture line and 1 to tearing of the esophagus above the suture line

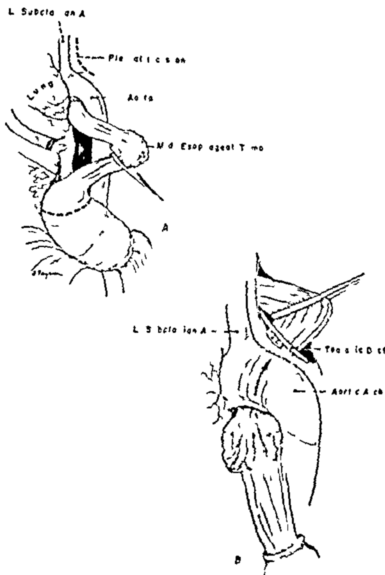


FIGURE 72 Operation for carcinoma of the mid-esophagus. In the first two drawings (A and B) the esophagus is mobilized behind the aortic arch and passed up and under. In drawings C and D the anastomosis is completed anterior to the aortic arch at a supra aortic level.

Superior mediastinal segment

Tumors lying above the aortic arch and extending to or through the thoracic inlet present the greatest technical difficulties in resection. In 9 patients with tumors in this location Sweet (41) was

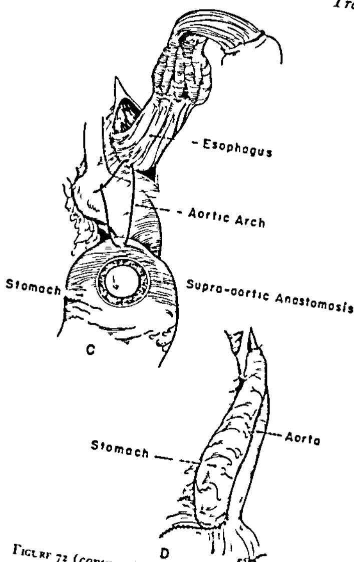


Figure 72 (*continued*) See legend page 244

able to resect the tumor in 6 with an operative mortality of 50 per cent None survived over 16 months

Transthoracic esophagogastrostomy In the conventional procedure the stomach and esophagus are mobilized through a left transthoracic approach as for a midthoracic lesion After careful maximum mobilization of the stomach has been achieved the organ is tacked to the mediastinal pleura and the fundus allowed

to lie free in the apex of the chest The thoracic incision is then closed the patient is turned on his back and redraped and the cervical portion of the esophagus is freed through the usual neck incision (Chapter 7) The sternal and medial clavicular inser

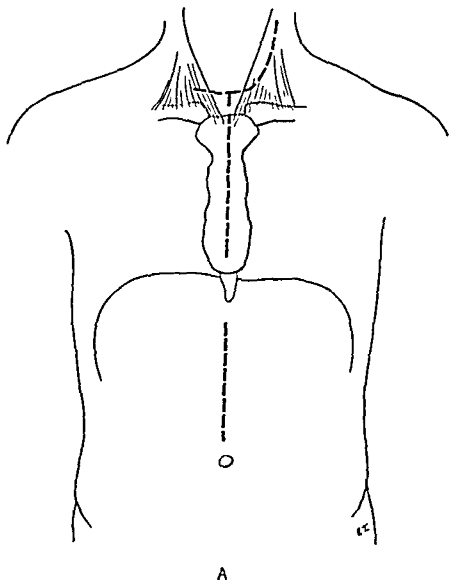


FIGURE [illegible] Waddell and Scannell's sternum splitting approach
(A) Skin incisions used for cervical mediastinal and abdominal approach to esophagus and stomach (B) The exposure as seen after division of the sternum and left innominate vein (C) The stomach passing through the anterior mediastinum and anastomosed to the uppermost portion of the *esophagus after esophagectomy* (See pages 248 249)

tions of the sternocleidomastoid are severed and retracted In some cases it may be advisable to resect the medial half of the clavicle and the left first rib and costal cartilage but this is not always necessary The pleura is then incised and the fundus delivered into the neck medial to the apex of the lung The anastomosis is carried out as previously described and the operative wound is closed in layers without drainage A complementary tracheostomy is usually advisable

Anterior sternum splitting approach Waddell and Scannell (47) have recently proposed an anterior sternum splitting incision for carcinoma of the lower cervical and superior mediastinal segments They have performed this in 6 patients and found that it provided excellent access to this difficult zone The entire procedure is carried out with the patient in the dorsal decubitus position (Fig 73A) Initially the cervical portion of the esophagus and the pharynx are exposed Lesions in this region can be inspected and mobilized with regional lymph nodes If the lesion can be resected the incision is extended vertically over the sternum and the latter split with a Lebsche knife A small rib spreader separates the halves of the sternum and exposes the upper mediastinum Division of the left innominate vein permits exposure of the esophagus to the level of the aortic arch (Fig 73B) The left recurrent laryngeal nerve is sacrificed and the esophagus is easily mobilized under direct vision Access to the upper mid esophagus can be facilitated if the aortic arch is gently lifted forward to the left In this manner the normal portion of the esophagus can be freed to the level of the carina with relative ease Beyond this the esophagus is freed by finger dissection Although occasionally individual vessels can be clamped and ligated it may be necessary to interrupt several small vessels without ligation We have seen only minimal bleeding at such a maneuver

As soon as the resectability of the lesion is established a second team begins mobilization of the entire stomach through a midline upper abdominal incision The freeing of the stomach is carried out so as to preserve the right gastric and right gastroepiploic

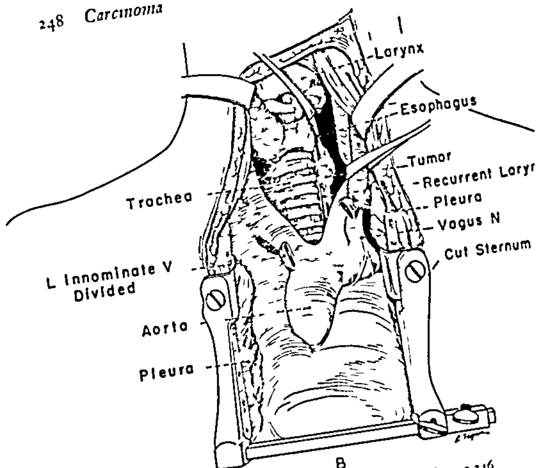


FIGURE 73 (*continued*) See legend page 246

vessels The short gastric and left gastroepiploic vessels are divided The left gastric artery is secured near its origin to preserve continuity between right and left gastric vessels along the lesser curvature The surgeon working from the abdominal approach mobilizes the lower esophagus and draws it down much as in the performance of a transabdominal vagotomy Through the hiatus the lower esophageal vessels can be pulled down with a finger and the proximal portions clamped and ligated Occasionally a vessel cannot be reached with instruments if so it is simply stripped out of the esophageal wall and allowed to retract In this manner the dissection from above meets the dissection from below The entire esophagus is thus freed and after severance from the stomach it is delivered into the superior mediastinum The mobilized

stomach is brought through an opening in the anterior portion of the diaphragm into the anterior mediastinum, the stomach will reach the pharynx by this route without undue tension. A three-layer anastomosis between the stomach and the pharynx or esophageal stump is made (Fig. 73C), fine interrupted silk sutures are used for this.

The abdominal wound is closed in layers with through-and-through stay sutures. The sternum is wired with heavy steel wire.

It is important to establish a tracheostomy before withdrawing the endotracheal tube. This can be placed to the right of the midline so that the tracheostomy does not communicate with the area of mediastinal dissection.

Colon Substitution. In an attempt to achieve an adequate resection for lesions in this inaccessible region we have attempted

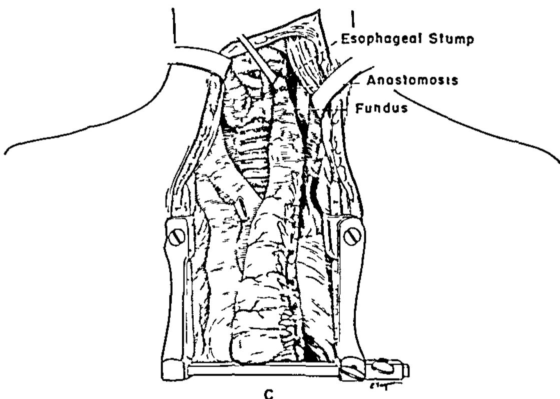


FIGURE 73 (*continued*). See legend page 46.

total esophagectomy utilizing the colon as an esophageal substitute (Fig 74) The technic of mobilizing and anastomosing the colon is identical to that described for lye strictures (Chapter 1.) A radical total esophagectomy can be carried out through the right side of the chest Since patients suffering from this

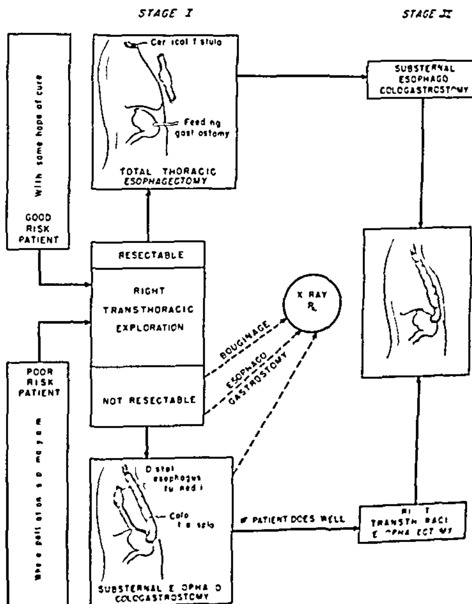


Figure 74 Various alternative procedures utilizing the colon as an esophageal substitute for high esophageal carcinomas.

disease are usually in the older age group and malnourished and frequently have complicating cardio-vascular-renal disease it may be advisable to stage such resections The substernal colon transplant can be done as the initial procedure without entering the chest Some assessment of operability can be made through the cervical incision If the patient tolerates the procedure well esophagectomy through the right side of the chest can be performed as a second stage If the lesion is found irresectable at the time of the first operation the cervical incision can be closed and the procedure terminated If dysphagia is severe the colon substitution can be completed and the second stage abandoned x ray therapy being substituted for the latter

The order of staging may be reversed by excising the esophagus at the first operation and performing a complementary gastrostomy and cervical esophagostomy At the time of the second procedure the colon is brought up to restore gastrointestinal continuity The gastrostomy may be closed at this time or closure may be delayed until the patient is again swallowing properly

Cervical portion of the esophagus

Carcinoma in this region is more commonly seen in women This type of tumor was first resected by Czerny (11) in 1877 the patient survived 15 months In 194 Wookey (51) described his procedure of cervical esophagectomy with skin flap closure which while not ideal continues to be the procedure of choice for localized carcinoma of the cervical portion of the esophagus The Wookey procedure is best suited to the small tumor that has not yet metastasized to regional nodes All other such cases are probably best treated by radiation or more radical surgery

The Wookey procedure is designed for cancer of the upper esophagus that is confined to the retrocricoid area The steps in the procedure are outlined in Figure 75 A large rectangular flap of skin including subcutaneous fat and platysma, is mobilized (Fig 75A) with its base at the posterior margin of the sternocleido-

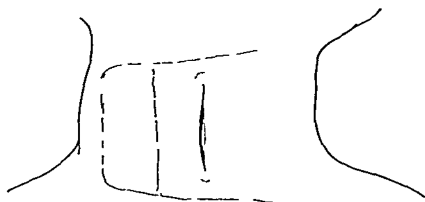
mastoid the base being about an inch wider than the free margin The flap is reflected back and protected with moist pads The omohyoid muscle is divided and the esophagus exposed If the growth is found to be resectable the sternal attachment of the sternocleidomastoid is divided and the lower half of the muscle is resected

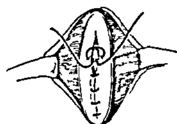
The sternohyoid and sternothyroid muscles and the thyroid lobe are removed care being taken to spare the recurrent laryngeal nerve The esophagus is mobilized and the growth resected The skin flap is next sutured into the defect (Fig 75B) The upper edge of the flap is then sutured to the pharynx and the lower edge to the esophagus as shown (Fig 75C) This is done in two layers approximating platysma to muscularis and skin to mucosa The free portion of the flap is turned back (Fig 75D) and the remaining defect is covered with a split thickness graft (Fig 75E) A complementary tracheotomy is carried out, this may be removed after a few days when a clear airway is present A Levin tube is inserted into the stomach through the esophagus for feeding purposes

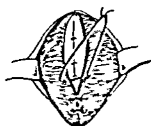
The second stage is carried out after complete healing has occurred — usually six to eight weeks later, this permits the flap to obtain a blood supply from its bed The steps for this stage are outlined in Figure 76 They consist of making an elliptical incision around the defect left at the first stage and approximating the inner edges to complete the tube and restore pharyngoesophageal continuity This inner layer is usually repaired with interrupted catgut sutures and covered by a second layer of platysma and a third layer of skin

When invasion of the larynx has occurred laryngectomy may be combined with esophagectomy at the time of the primary procedure This is a mutilating operation with resultant loss of voice and it should not be attempted unless there is no evidence of spread and the lesion seems to be curable

Mustard and Ibberson (3) found 3 five year survivors among 45 patients with carcinoma of the hypopharynx and cervical esoph







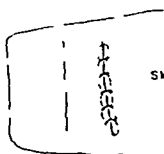


FIGURE 76 Wookey procedure second stage An elliptical incision is mad surrounding the skin opening and the latter is closed in layers.

agus who were treated surgically Sweet reported (41) 17 cases of cervical carcinoma, in 10 of which resection was possible Seven patients had died of their disease, 2 were surviving at 18 months, and 1 had undergone operation only recently

It is our belief that the Wookey procedure is applicable to only very few patients with cervical neoplasms and even in those cases permits only a limited resection We are currently attempting a more radical and extensive cervicothoracic approach in all such cases utilizing either the stomach or the colon (Chapter 1[illegible]) as esophageal substitutes Our experience is still too limited to permit any conclusions

Prognosis

In considering surgery for carcinoma of the esophagus a sharp distinction must be made between palliative resections and those undertaken for curative purposes Garlock and Klein (14) excluding operative deaths reported a [illegible] per cent over-all five-year survival rate in patients operated on for cancer of the esophagus They emphasize the important influence of lymph node involvement on the survival rate The majority of late survivors showed no lymph node spread

After excluding palliative procedures Sweet (42) reports a 14 per cent five-year survival for carcinoma of the midthoracic segment and a 34 per cent five year survival for cancers of the lower portion He also emphasizes the importance of negative nodes for a good prognosis

Nakayama *et al* (33) have reviewed their experience with 410 patients over a ten year period Resectability amounted to 41 3 per cent and the mortality rate was 4 7 per cent They found 69 cases of five year survival in the world literature including 19 of their own

In a review of 381 cases of carcinoma of the esophagus admitted to the Toronto General Hospital during the period [illegible] 1953 inclusive Mustard and Ibberson (3[illegible]) found only 1 patient

with squamous cell carcinoma of the thoracic portion of the esophagus who survived 5 years after surgery. Of the whole series only 8 patients (3.0 per cent) survived 5 years; 4 of these cases were hypopharyngeal lesions and 2 were adenocarcinomas of the abdominal portion of the esophagus. One was treated by radiation. Female patients have a slight advantage over males in expectation of survival.

Carey and Clagett (9) found that the results of resection were better in patients with squamous-cell tumors of the lower esophagus than in those with adenocarcinoma of the cardia. Sweet (42) noted no significant difference in the prognoses of these two conditions. Shedd, Crowley, and Lindskog (38) reported 3 five-year survivors in 30 patients undergoing esophageal resection, all had adenocarcinomas. No patient with epidermoid carcinoma survived longer than 27 months.

Recent studies by Gephart (15) at the Massachusetts General Hospital suggest that the so-called 'sensitization reaction' described by Graham and Graham (17) in carcinoma of the cervix may also be of value in selecting those squamous cell tumors of the esophagus which will respond well to x-ray therapy. Preliminary results suggest that most esophageal carcinomas are not responsive to radiation therapy. By the standards accepted as an index of radiation sensitivity for carcinoma of the uterine cervix very few esophageal carcinomas would seem to be sensitive to radiation. It is possible that different criteria may be necessary for selecting esophageal carcinomas from those used for carcinoma of the cervix. The eventual findings of this study should be of considerable therapeutic and prognostic value.

Sweet's (42) editorial on the present status of treatment of carcinoma of the esophagus should be studied carefully by any physician who undertakes the care of malignant lesions of the esophagus.

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Sarcoma

20

A sarcoma is a malignant tumor arising from connective tissue.

Incidence

Sarcoma of the esophagus is very rare. In all the clinical and autopsy records of malignant disease of the esophagus in the Massachusetts General Hospital only 4 cases of sarcoma of the esophagus could be found. According to Jackson (4) sarcoma occurred in 7 out of 935 cases of malignant disease of the esophagus.

Chapman (7) was the first to report (1877) a case of sarcoma of the upper esophagus. The history was that of a 45 year old woman who complained of food, especially meat, sticking in her throat with pain in the throat, head and neck. There was also occasional blood-streaked, thick, whitish yellow sputum. At esophageal exploration the bougie was checked by a hard, resistant surface. The diagnosis was established by microscopic examination of the sputum, which revealed many long spindle cells.

The next cases were reported by Targett (46) in 1888 and by

Shaw (43) in 1891 In Shaw s case there was an ulcer encircling the esophagus and perforating the posterior wall of the trachea a short distance above its bifurcation In 1893 Rolleston (41) reported a case of sarcoma of the esophagus with secondary growth to the bone In his case autopsy showed the growth to be adherent to the pericardium and to the right lung Toward the upper part of the stricture there was a fistulous passage through the growth into a gangrenous cavity in the lower lobe of the right lung Other cases were reported by Ogle (35) and James (26) In 1900 Starck (45) collected 7 cases and added 2 more In 1902 Howard (21) reported a case of primary sarcoma of the esophagus and concluded that in most of such cases the tumor was located in the lower part of the esophagus was more common in males than in females and unlike carcinoma might appear early in life from the age of 4 up to the age of 25 He believed that the course was likely to be more rapid and that a fatal issue was to be looked for earlier than in carcinoma

Hacker (18) collected 21 cases in 1909 Hacker and Lotheissen (19) stated in 19 6 that only 38 cases had been described and in some of these the origin was doubtful According to Dvorak (10) Simon (44) said in 19 8 that there were only 30 objection free cases in the literature Guisez (17) reported a case of sarcoma of the esophagus in 1927 the first biopsy showing questionable sarcoma after which radium was applied and improvement resulted The second biopsy 6 months later showed carcinomatous change in a sarcoma Guisez believes that sarcomas are more painful than carcinomas and that pain is likely to precede the dysphagia In 1931 Dvorak (10) described a polypoid rhabdomyosarcoma in the cervical esophagus which contained muscle cells of the striated type spindle cells giant cells round cells and fibrous connective tissue He says that there are two types of sarcoma (1) polypoid which contains spindle cells and which is relatively benign and rarely metastasizes and (2) the diffuse infiltrating type which contains round cells and early gives rise to distant metastases He says the pain in sarcoma is often early and

severe, unlike that of carcinoma He collected 31 cases from the literature

In 1935 there was a Massachusetts General Hospital Cabot Case report (6) of a leiomyosarcoma *of the distal third of the* esophagus In 1935 Jaleski and Waldo (25) reported a case of *primary melanotic sarcoma of the esophagus undiagnosed before* autopsy with no symptoms of esophageal obstruction before death They found only 3 other similar cases in the literature (1, 27 49) In 1937 Menne and Birge (33) reported a case of primary leiomyosarcoma of the upper third of the esophagus which grew slowly and invaded slowly Its extension into the trachea anteriorly probably resulted in implantation metastases in the lungs *In 1940* Pearlman *(36) referred to 23* cases *of* so called carcinosarcoma of the esophagus and added 8 cases of his own which *might have* been termed "carcinosarcoma but were *prob* ably cases of anaplastic squamous-cell carcinoma with transi tional-cell features Pearlman believes that most of the so called carcinosarcomas of the esophagus are really primary carcinomas In 1941 French and Garland (15) reported what they called the fifth case of leiomyosarcoma of the esophagus In 194 Pennes (37) reported the sixth case of leiomyosarcoma of the esophagus involving the middle and lower third, the tumor extended into the mediastinum simulating a primary mediastinal tumor The patient *did not complain of* dysphagia when seen In 1949 Clark (8) re ported what was probably the first successful resection of an *esophageal sarcoma In 1948* Resano *and* Albanese (38) reported a case of primary sarcoma of the esophagus In 1950 Fortat Jacob and Badaro (30) reported two *primary pedunculated* sar comas of the esophagus Another case was reported by Mauro and De Escobar (32) in 1951

Ishikawa and Nakagawa (23) in 1956 reviewed the literature on esophageal sarcoma finding in all *64* cases to which they added one of their own

Pathology

Many of the reports in the literature do not specify the type of sarcoma Excluding so called carcinosarcoma (which is probably carcinoma) the following types of sarcoma have been reported as occurring in the esophagus

Round cell sarcoma has been reported by Howard (1), Bertholet (4), Rolleston (41), Starck (. cases) (45) Wegener (50), and Huismans (2.) According to Dvorak (10) Ewing (11) said *that many round cell sarcomas are nothing less than anaplastic carcinomas*, and Fischer (1.) stated that round cell tumors are probably either lymphosarcomas or embryonal—that is, undifferentiated—carcinomas

Spindle cell sarcomas have been reported by Chapman (7), Targett (46), Ogle (35), James (.6) Hofmann (.0), Livingood (.8) and Notthaft (34) According to Dvorak (10) tumors containing some spindle cells either large or small and some giant cells were described by Borrmann (5) Donath (9), von Licken (48), Frangenheim (14) Hofmann (20) Notthaft (34) Ricke (39), and Schmincke (42)

Melanotic sarcoma has been reported by Jaleski and Waldo (25) and Baur (1) According to Jaleski and Waldo 3 other similar cases are reported in the literature

In 195. Garfinkle and Cahan (16) presented a case of melanocarcinoma of the esophagus with metastases to the paratracheal lymph nodes Fowler (13) also in 195. added a case of malignant melanoma of the esophagus Robertson (40) added another case of malignant melanoma of the esophagus with metastases to regional lymph nodes lung liver pancreas and peritoneum Loring and Zeppa (.9) in 1956 found 16 cases of melanocarcinoma of the esophagus in the world literature since 1906 they described the seventeenth case

Leiomyosarcoma seems to be about as rare as melanotic sarcoma having been reported by Hacker (19) Baur (1), Menne

and Birge (33) French and Garland (15), and Pennes (37), and in a Cabot Case report (6)

Rhabdomyosarcoma appears to be even more rare than the other types The case summarized by Benedict (3) was the fourth *reported case of rhabdomyosarcoma* Metastases were not demonstrated in Dvorak's case or in the case reported by Benedict Two further cases of rhabdomyosarcoma have come to our attention as reported by Thorek and Neiman (47) and by Mainero (31)

According to Dvorak (10), polypoid spindle-cell sarcomas grow slowly and destroy little tissue The infiltrating and ulcerating round cell type can quickly reach a great dimension and tend to obstruct and close the lumen, compress the trachea, and cause edema of the larynx There may be penetration to contiguous organs purulent pleurisy, pulmonary gangrene and eventual esophageal perforation Metastases are most likely to occur in the diffuse infiltrating type of round cell sarcoma They occur early and are widely distributed to lymph nodes, inner organs and bones

Symptoms

The symptoms closely resemble those of carcinoma Most writers agree however that sarcoma of the esophagus usually occurs at an earlier age than does carcinoma and that pain is more likely to be present in sarcoma Dysphagia may or may not be present It is likely to be present when there is marked proliferation or ulceration with stenosis Sometimes the course is entirely symptomless and the lesion is discovered only at autopsy Cachexia begins earlier than in carcinoma probably because pain interferes with sleep and leads to loss of appetite and emaciation which according to von Eicken (48) is greater than that which corresponds to the stenosis

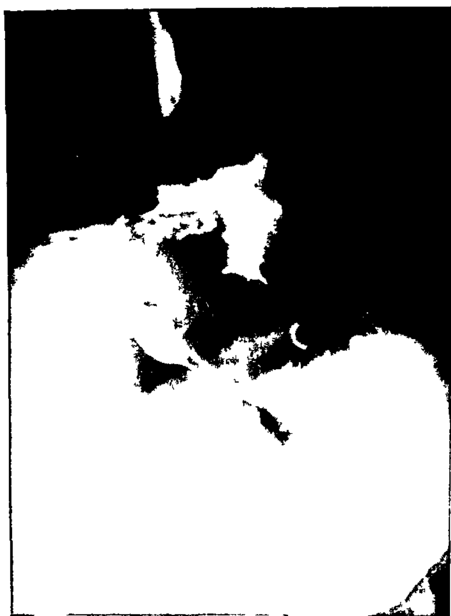


FIGURE 7. X ray appearance of large leiomyosarcoma of lower third of the esophagus with perforation simulating carcinoma in a herniated stomach

Diagnosis

History and x-ray findings (Fig 77) are similar to what is found in carcinoma of the esophagus (pages 228 229) The only methods for making an exact preoperative diagnosis are esophagoscopy and biopsy The two sarcomas which we have seen by esophagoscopy were both nodular and were grossly indistinguishable from carcinoma The diagnosis was correctly made by histologic examination of the esophagoscopic biopsy specimen

Treatment

The treatment of esophageal sarcoma is radical resection if possible The technic is identical to that used for carcinoma (Chapter 19) This is of considerable benefit in the lymphosarcomatous (Hodgkin's) type but of comparatively little benefit in the fibrosarcomas

Prognosis

Unless the neoplasm can be radically removed by surgery the prognosis is bad Bayer (2) however in 1941 reported a case of sarcoma of the esophagus free of symptoms 15 months after cure by roentgen irradiation

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Lymphoma

21

According to Gall and Mallory (3) many generic terms have been utilized to designate those maladies of the lymphatic system which are characterized clinically by progressive tumor-like enlargement of lymphoid tissue with eventual death and are characterized histologically by multiplication of one or more of the elements normally present in lymph nodes to the point of destruction of the nodal architecture. Of these, "malignant lymphoma" seems to have won most general usage—in this country at least—and has the advantage of being noncommittal as to pathogenesis.

Incidence

Primary lymphoma of the esophagus is exceedingly rare. Cases which have been reported as lymphosarcoma probably belong in the lymphoma group. Although all types of malignant lymphoma have occurred in every age group, it is more frequently seen between the ages of 50 and 60, except for Hodgkin's disease, which occurs with greater frequency between the ages of 30 and 40 and is not uncommonly found in youth before the age of 20. At the other extreme of age, clasmatocytic and follicular lymphomas have the greater incidence, with only about 20 per cent occurring

before the age of 40 There is a preponderance of males over females in a ratio of [illegible]-3 to 1, depending on the type of malignant lymphoma

Stephan (5) in 1889 reported the case of a lymphosarcoma in a 4 year-old boy this is the lowest age so far recorded Jackson (4) reported the case of a 16-year-old boy with lymphosarcoma involving the anterior wall of the esophagus at the level of the crossing of the left main bronchus The patient had no dysphagia but there was a bronchoesophageal fistula

Weyde (6) described a 23-year-old man who was suffering from Hodgkin s disease After six and one half years illness he noted difficulty in swallowing, the origin of which proved to be an infiltration 8 cm in length situated just above the cardia Esophagoscopy revealed polypoid grayish red soft tumors which bled readily when touched macroscopically they could not be differentiated from carcinoma Roentgen examination also showed changes resembling those in carcinoma The microscopic examination however established the diagnosis of Hodgkin s disease After the termination of roentgen treatment with a tumor dose of [illegible]864r increased liquefaction of the granulation tissue in the wall of the esophagus was observed Four weeks later the outlines were even and variable with only slight stiffness in the previously infiltrated areas Bichel ([illegible]) also reported a case of Hodgkin s disease in which there was marked esophageal involvement with only slight dysphagia and in which the benefit obtained from irradiation therapy was very impressive

Etiology

The etiology of lymphoma is unknown

Pathology

Gall and Mallory (3) classify malignant lymphoma into seven types as follows

(1) *Stem cell lymphoma* The lymphoid stem cell is a large

undifferentiated cell of mesodermal origin which develops the ability to differentiate into various forms of blood cells Cells of this type constitute the predominant element in stem cell lymphomas

(2) *Clasmatocytic lymphoma* The cells in these tumors simulate more or less normal clasmatocytes or monocytes

(3) *Lymphoblastic lymphoma* The predominating cell in this lesion is a lymphoblast

(4) *Lymphocytic lymphoma* The predominating cell in this lesion is indistinguishable from a normal lymphocyte

(5) *Hodgkin's lymphoma* Hodgkin's disease constitutes the commonest and most readily recognizable lesion among the lymphomas Hodgkin's disease is essentially polycellular and contains granulocytes plasma cells, clasmatocytes and fibroblasts but the pathognomonic elements which they contain are stem cells which tend to develop large multilobed or multinucleated forms

(6) *Hodgkin's sarcoma* Hodgkin's sarcoma retains the fundamental background of Hodgkin's lymphoma the basic cell being the tumor stem cell Multinucleated cells predominate and mitotic figures are very numerous

(7) *Follicular lymphoma* Follicular lymphoma manifests itself through complete replacement of normal lymph node architecture by multiple follicle-like nodules of varied size and approximation

Our own experience is as follows

Case 1 In the case of proven malignant lymphoma of the esophagus reported by Benedict (1) (Fig 78) the esophagus was resected and the pathologic diagnosis was malignant lymphoma lymphoblastic type It was primarily located in the submucosa and seemed to infiltrate the muscularis and the deeper layers of the mucosa

Case 2 Since the above mentioned case was reported another example of malignant lymphoma has come to our attention that of a 19 year old boy who entered the hospital with a chief complaint of difficulty in swallowing considerable weight loss and



FIGURE 78 X ray showing lymphoblastoma in the middle third of the esophagus This probably arose however from mediastinal lymph glands The diagnosis was established by lymph gland biopsy In 1935 the mediastinal involvement responded to x ray treatment In April 1938 there was evidence of involvement of the lung parenchyma In October 1938 there was onset of dysphagia In December 1938 the patient was able to swallow liquids only with difficulty The above x ray shows a mass involving the esophagus producing definite obstruction to the flow of barium After further x ray treatment the involvement of the esophagus disappeared almost completely and the difficulty in swallowing subsided almost entirely

fatigue Cervical and axillary lymph nodes were palpable and a biopsy of one of these was reported as Hodgkin's disease X ray examination showed an extrinsic para- and retro esophageal mass with ulceration of a considerable portion of the upper segment of the esophagus Esophagoscopy disclosed a firm annular nodular, easily bleeding, ulcerating lesion of the upper esophagus which had more the appearance of an intrinsic lesion of the esophagus than of extrinsic invasion Bouginage was carried out and a biopsy specimen obtained which was reported consistent with irradiated Hodgkin's disease (the patient had previously had x-ray treatment at another hospital) Further x ray treatment was advised this was given at another hospital It is usually difficult or impossible to determine in which organ malignant lymphoma originates In this case dysphagia was the chief complaint when the patient was admitted to the Massachusetts General Hospital The fact that it was evident at the time of esophagoscopy suggested primary involvement of the esophagus

Case 3 Still another case is of interest—that of a 5 year old man who entered the hospital because of difficulty in swallowing of six weeks duration In this case x-ray examination demonstrated a huge tracheobronchoesophageal fistula Esophagoscopy revealed marked narrowing of the esophageal lumen Several large biopsy specimens were obtained these were reported as undifferentiated carcinoma The patient failed rapidly and died Autopsy showed malignant lymphoma clasmatocytic type with involvement of bronchial mediastinal and abdominal lymph nodes and the right lung Esophagopulmonobronchial fistula with multiple abscesses also was demonstrated It is interesting to note that the pathologist may not be able to differentiate lymphoma and undifferentiated carcinoma on the basis of a biopsy specimen

Symptoms

The symptoms produced by lymphoma depend entirely upon the location of the disease When the tumor occurs in the esoph

agus it is likely to produce dysphagia, but there are so few well-documented cases of lymphoma of the esophagus that no specific symptoms can be described. Undoubtedly lymphoma which involves the mediastinum but is primary elsewhere in the body may infiltrate the wall of the esophagus and produce extrinsic pressure on the organ without giving rise to characteristic symptoms unless there is enough infiltration or extrinsic pressure to produce narrowing and dysphagia.

Diagnosis

The diagnosis of esophageal lymphoma is exceedingly difficult. There is usually a history of dysphagia and there may be a history of lymphoma involving other parts of the body. X-ray examination may reveal a mass in the esophageal lumen if the disease is primary in the esophagus or may disclose only extrinsic pressure. In any case esophagoscopy should be performed to determine whether a positive diagnosis can be made by biopsy. Bouginage may be carried out at the same time.

Treatment

If the disease appears to be localized to the esophagus surgical resection may be attempted. On the other hand x-ray treatment is usually effective and certainly should be used as the treatment of choice. The addition of nitrogen mustards and other similar compounds now in a developmental phase promises even greater effectiveness for radiation treatment.

We operated on a young man who had responded well to radiation therapy for lymphoma but developed a tracheoesophageal fistula; the fistula had caused repeated bouts of aspiration pneumonia. An attempt to repair the fistula and restore gastrointestinal continuity by means of a colon transplant was made but the patient succumbed after operation to pulmonary complications. In performing thoracotomies on such patients the surgeon

must remember the pulmonary limitations imposed by tumor infiltration radiation damage and fibrosis secondary to aspiration when a tracheoesophageal fistula is present

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Metastatic Tumors

22

Metastatic tumor involving the esophagus is exceedingly rare. The earliest report is that published by von Recklinghausen (6) in 1885 of a tumor of the tibia diagnosed as myxochondrosarcoma with a pea sized nodule in the muscularis of the esophagus. In 1895 Spiegelberg (4) reported multiple nodules in the esophageal mucosa which had metastasized from melanosarcoma of the eye. In 1942 Gross and Freedman ([illegible]) reviewed the literature including the above mentioned cases and added one case of their own in which carcinoma had metastasized to the esophagus from a carcinoma of the prostate. In their case the primary carcinoma of the prostate was silent but the metastatic lesion encircled the wall of the esophagus without increasing its external diameter, the mucosa was intact and normal over the mass which was 3 cm long and nearly obliterated the lumen. In their review of the literature Gross and Freedman found records of 4[illegible] cases of secondary tumor of the esophagus in most of which there was no esophageal obstruction.

In 1944 Toreson (5) reviewed 3048 consecutive autopsies in adults and found 599 cases of cancer in 19 of which there was secondary involvement of the esophagus—an incidence of 3[illegible] per cent. He also found 7 additional cases of secondary tumor of

the esophagus, which brought the total to 26 cases. Of these 24 were carcinomas, 1 was a lymphosarcoma, and 1 probably a melanoma. The organs in which the tumor originated and the numbers for each were as follows: trachea 8, stomach 7, larynx 4, breast 2, pancreas 1, testes 1, metastatic lymph nodes 1, origin undetermined 1. Most cases of metastatic tumors are the result of direct invasion from neighboring organs or from mediastinal glands. However, in 2 of the breast cases described by Toreson a metastatic tumor nodule was found in the wall of the esophagus, but there was no involvement of the mediastinal glands. Involvement of the esophagus from the stomach usually occurs by direct extension, but in 1 case a primary carcinoma of the stomach originating in the pylorus had not invaded the remainder of the stomach but did involve the esophagus. In 12 of Toreson's collected cases sufficient esophageal obstruction occurred to cause clinical symptoms. In somewhat fewer than 50 per cent of the cases there was more or less severe dysphagia.

In 1941 Ranieri (3) described an unusual case of cutaneous melanosarcoma with probable metastases to the esophagus but no stenosis.

In 1953 (1) an oat-cell carcinoma of the left lung was reported with metastases to mediastinal, azygos, and cardiac lymph nodes, some of which had ulcerated into the esophagus and produced obstruction.

The surgeon must keep in mind the possibility of secondary involvement of the esophagus as a cause of dysphagia. It is of little benefit to the patient to resect such a lesion in the belief that it is a primary neoplasm. More commonly the esophagus will be involved by direct extension from tumors of the stomach, the liver, or the pancreas. In such cases operability will depend on the characteristics and the extent of the primary tumor.

Palliative x-ray treatment should be considered, as well as bougienage, but the prognosis is bad.

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FOREIGN BODIES IN THE ESOPHAGUS

23

Any bolus of food or any other object which becomes lodged in the esophagus is known as a foreign body.

ETIOLOGY

The various contributing factors of foreign body in the esophagus are as follows:

Age. Infants and young children are likely to put into their mouths everything that they can reach. Many of these objects are accidentally swallowed, and those that are too large or too sharp to pass become lodged in the esophagus. Old people may be edentulous or may have poor teeth and hence masticate poorly. In such persons foreign bodies are likely to get stuck in the esophagus.

Carelessness. This is undoubtedly the commonest cause of foreign body in the esophagus. Incomplete mastication may result in the swallowing of a large bolus of food which may become stuck in the esophagus. Also because of incomplete mastication

pieces of bone or other foreign material may inadvertently be swallowed and become lodged in the esophagus Improperly fitting dentures result in lack of mastication, which in turn leads to the possibility of foreign body The denture itself may be swallowed and become a foreign body (Fig 79, Plate IX, #10) Eating in an intoxicated state may lead to the aspiration or swallowing of a foreign body

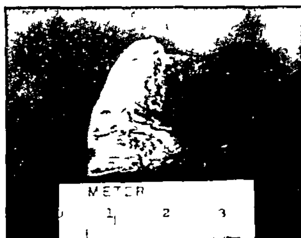


FIGURE 79 This large dental plate which had been stuck in the lower esophagus was successfully removed by esophagoscopy using rotation forceps A corner of this plate projected beyond the barium column in such a way that it was mistaken by the radiologist for peptic ulcer of the esophagus

Type of foreign body Obviously, the size, shape, and nature of the foreign body determines whether or not it passes smoothly or becomes lodged in the esophagus

Abnormality of the esophagus Although many foreign bodies become lodged in the normal esophagus the hazard is greatly increased by any abnormality such as stricture, tumor, web achalasia, spasm or paralysis

PATHOLOGY

A foreign body may become lodged at any level in the esophagus However there are certain sites of predilection, namely, just

below the cricopharyngeus and just above the cardiac orifice At the site of lodgment the esophageal wall very soon becomes irritated red and edematous Sharp foreign bodies are likely to perforate early if not promptly removed Large irregular or jagged foreign bodies also may cause perforation If no treatment is instituted periesophagitis, mediastinitis and mediastinal abscess are likely to develop rapidly, with fulminating symptoms and rapidly fatal termination However, skillful management, with bed rest nothing taken by mouth antibiotics intravenously administered fluids etc, may today result in saving the lives of many patients with mediastinitis who formerly would have died

Symptoms

Choking gagging, or coughing while eating are common symptoms which should make one suspect foreign body There also may be a history of having swallowed too large a mouthful or of having felt a hard or sharp object in the back of the throat and feeling as if something had stuck in the esophagus Pain is frequently present especially with sharp objects The pain may be sharp and fairly constant or may be noted only at the time of deglutition Dysphagia is usually present even the swallowing of liquids may be painful Sometimes the foreign body passes on into the stomach but the pain persists in the esophagus In such a case if the foreign body is nonopaque esophagoscopy is the only means of determining whether it is still present in the esophagus

Complications

Foreign bodies that have been in the esophagus for a long time cause irritation with infection esophagitis periesophagitis mediastinitis and possibly mediastinal abscess Sharp foreign bodies in the esophagus may perforate early after only a short sojourn The treatment of these serious complications means careful treat

ment of the patient as a whole nothing should be given by mouth adequate fluids should be administered intravenously, antibiotics should be given intramuscularly and a thoracic surgeon should be called in consultation

Diagnosis

A history that suggests foreign body is very important in making the diagnosis However a foreign body may be present when there is very little history to indicate it and conversely a foreign body may be absent even when there is a history that suggests it Unless complications have occurred such as perforation with mediastinitis or abscess physical examination is non contributory Radio opaque foreign bodies are readily demonstrable by x-ray examination, in fact they can usually be seen at fluoroscopy and it must not be forgotten that they may have lodged anywhere from the upper esophagus to the rectum Thin pieces of bone are almost never visible by x ray

Giving the patient a swallow of barium is usually inadvisable since it often contributes nothing and gives the physician a false sense of security In that connection we know of a patient treated at another hospital who had definitely swallowed a pork bone (Fig 80) but was given a barium swallow with negative results and was allowed to go home for a period of four days during which time the bone perforated the esophagus and mediastinitis developed (Fig 81) The patient finally recovered after esophagoscopic removal of the foreign body with surgical drainage and a stormy rather long convalescence Furthermore the administration of barium in a questionable case of foreign body makes subsequent esophagoscopic removal much more difficult by obscuring the field with barium this also applies to barium capsules Quite often the only method of making a positive diagnosis is esophagoscopy It is far better to perform an esophagoscopy and find no foreign body than to omit esophagoscopy and leave a foreign body in the esophagus

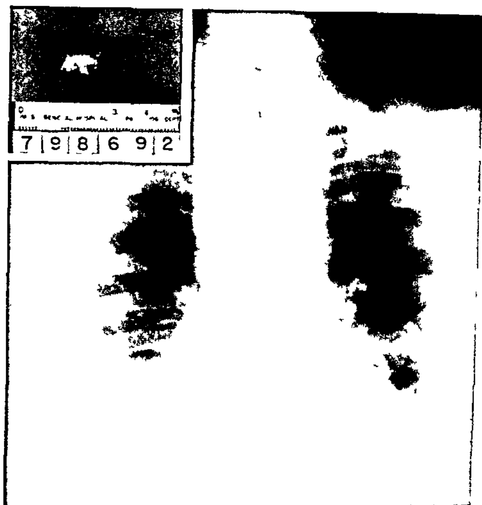


Figure 80 Jagged appearance of pork bone removed by esophagoscopy after 4 day sojourn during which time it caused a spontaneous perforation of the esophagus See the text (page 85) for the inexcusable reason for delay

Figure 81 X ray appearance of mediastinitis caused by pork bone in the esophagus (see Figure 80)

Treatment

Esophagoscopy

The treatment for foreign body in the esophagus obviously is removal Occasionally when the history is vague and the x ray examination non contributory it is reasonable to delay esophagoscopy If however, there is a fairly definite history esoph

agoscopy should be undertaken promptly In adults it can usually be performed with proper sedation under local anesthesia However in a nervous patient or in the case of a difficult foreign body, complete relaxation is advisable it may easily be obtained with intravenous administration of pentothal anectine Although foreign body in children can sometimes be removed without anesthesia modern anesthesiology is so easy and safe that it seems unnecessarily cruel to subject a child to the mental trauma and physical discomfort of esophagoscopy without anesthesia Moreover the procedure is easier for the esophagoscopist if the child is absolutely quiet and asleep We therefore recommend intratracheal ether with oxygen as needed believing that with a tracheal tube of proper size there is very little risk of laryngeal edema If symptoms of laryngeal edema develop such as cough stridor wheeze, or dyspnea the use of a high humidity tent for a few days will usually cause the symptoms to subside On rare occasions a tracheotomy may be necessary

It must not be forgotten that foreign bodies thought to be in the esophagus may occasionally prove to be in the tracheobronchial tree and *vice versa* Blind attempts to force foreign bodies into the stomach are to be condemned as they are exceedingly dangerous Unless perforation has already occurred most foreign bodies in the esophagus can safely be removed through the esophagoscope If there is reason to believe that there has been perforation into the neck or mediastinum a direct surgical approach may be more advisable than esophagoscopy Each object presents its own problem of esophagoscopic removal Skill should be acquired through actual removal of various types of foreign bodies in animals Some of the foreign bodies encountered and the methods of their removal are as follows

(1) *Bolus of meat* In some cases a bolus of meat may be successfully dissolved by the use of Caroid a vegetable digestive ferment which is made from papaya The powder should first be made into a paste with water and then into a solution which the patient sips (1 heaping teaspoonful of powder to 6 oz of water)

If this is not rapidly successful, esophagoscopy should be undertaken The bolus is usually found at the lower end of the esophagus and ordinarily can be readily removed with large ball forceps

(2) *Bones* These are of varying shapes and sizes (Fig 82, see also Plate IX, #11) Flat pieces of bone should be grasped with the rotation forceps, with sharp points trailing in so far as possible, and with the plane of the bone in the coronal plane of the body so that when the forceps bring it to the level of the cricopharyngeus the action of that muscle will not strip it off (This plane is exactly at right angles to the plane in which flat objects should be removed from the bronchus in order to bring them

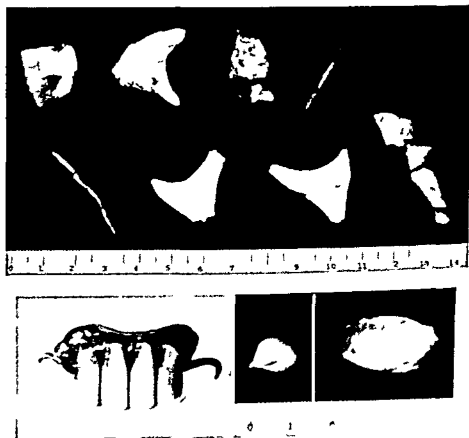


FIGURE 82 Chicken and fish bones of various sizes and shapes which have been stuck in the esophagus and successfully removed by esophagoscopy Note that many of them have sharp jagged points and edges. A flat bone should be grasped with rotation forceps with the points trailing

past the vocal cords) Sharp pieces of bone can usually be grasped with the side curved or ball forceps the point carefully disengaged from the esophageal mucosa, and the bone fragment brought *into the lumen of the esophagoscope if possible Foreign bodies* which are too large to be brought within the lumen of the esophagoscope are brought up to the lip of the tube and held firmly there while the instrument and the forceps are removed together with the foreign body It is in such cases that stripping at the cricopharyngeus must be carefully avoided by having a firm grasp on the foreign body and by having the foreign body in the right plane

(3) *Coins and other flat objects* These can usually be grasped with the rotation forceps brought to the mouth of the tube, rotated into the proper plane to avoid stripping at the cricopharyngeus and readily removed (Plate IX, #9 #1.)

(4) *Open safety pins* Unfortunately for the patient and also for the endoscopist, most safety pins are swallowed open They enter the esophagus with the spring down and the point up making removal rather difficult in many cases Many methods of removal have been described, depending on the size, condition and location of the pin In the unusual event that the spring of the pin is upwards and the point downwards, the spring is grasped with the rotation forceps and the pin is automatically closed as it is drawn into the mouth of the esophagoscope Even when the point is up very small pins may be grasped by the spring and the pin tipped over this makes it point downward and renders removal easy Since trailing points do not perforate it is often possible to take a rather large safety pin with the point up grasp the spring with rotation forceps push it gently downwards into the stomach where it can be made to dangle and turn itself around and then withdraw into the esophagoscope Another valuable method is to sheathe the point of the pin by grasping it with the tack and pin forceps bringing it well into the tube mouth and leaving the keeper outside the esophagoscope the idea being that the rounded blunt keeper will cause no damage during re

moval In grasping the point of a pin which has become slightly embedded in the wall of the esophagus, it is important to remember that it should be disengaged gently, and that forceps space may be gained by pressing against the wall of the esophagus with the lip of the esophagoscope

Surgery

Occasionally a pin or other sharp object may be seen by x ray to be in such a position that it either has already perforated the esophagus or otherwise presents an almost hopelessly difficult problem for the endoscopist In such cases surgery should be performed without preliminary endoscopic attempts at removal (Fig 83)

In infants an esophageal foreign body should suggest the possibility of pre-existing esophageal disease, such as a congenital



Figure 83 This open safety pin could not be safely removed with the esophagoscope Thoracotomy was required

narrowing Gross (1) found such an abnormality in 10 per cent of a series of 151 infants with esophageal foreign body

When endoscopic extraction is unsuccessful or is contraindicated the surgical approach depends on the location of the foreign body in the esophagus, it will be made through either the left side of the neck or the left or right thorax as described in Chapter 7 Once the esophagus is mobilized a longitudinal incision is made into the lumen of the esophagus and the foreign body extracted The esophagotomy is closed with interrupted silk sutures the first layer in the mucosa with knots tied within the lumen and a second layer in the muscularis Thoracotomy incisions are drained with a catheter cervical incisions are not drained Systemic antibiotics are administered before and after operation

Prognosis

The prognosis depends on the nature of the foreign body the length of its sojourn in the esophagus and the skill of the endoscopist

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Extrinsic Pressure

24

Any mass which presses on any part of the esophagus may give rise to esophageal symptoms such as dysphagia, regurgitation or vomiting

Etiology

In the neck tumors of the thyroid parathyroid carotid body (9) or thymus glands sometimes press upon or even invade the esophagus Cervical exostoses may on rare occasions cause dysphagia

In the mediastinum extrinsic pressure on the esophagus may be caused by intrathoracic goiter, mediastinal tumor mediastinal granulomas (including tuberculosis and sarcoid) carcinoma of the bronchus pressing upon or invading the esophagus dilated left auricle vascular anomalies (e g , dysphagia lusoria) and aneurysm

Incidence

Lesions of the neck

Thyroid (Fig 84) A parathyroid adenoma causing deviation of the esophagus was discussed in 1951 in the case records of the

Massachusetts General Hospital (1) Cope (2) has reported that 3 out of 96 parathyroid adenomas showed x ray evidence of extrinsic pressure on the esophagus Cope *et al* state that in rare cases carcinoma of the parathyroid may invade the esophagus, necessitating the removal of part of that organ (3)



Figure 84 A 43 year old woman complained of fever malaise sore throat and difficult painful swallowing of 6 months duration X ray (above) revealed a soft tissue mass in the left lower neck compressing the esophagus and displacing it toward the right Total thyroidectomy was performed for adenocarcinoma grade III which had invaded and ulcerated the esophagus The patient did well after the operation

Thymoma (Fig 85)

Exostoses Exostoses are quite common but rarely cause dysphagia Kertzner and Madden (8) however reported a case of cervical exostoses which actually did cause dysphagia Exostoses also are of significance because of the danger of esophageal perforation during esophagoscopy

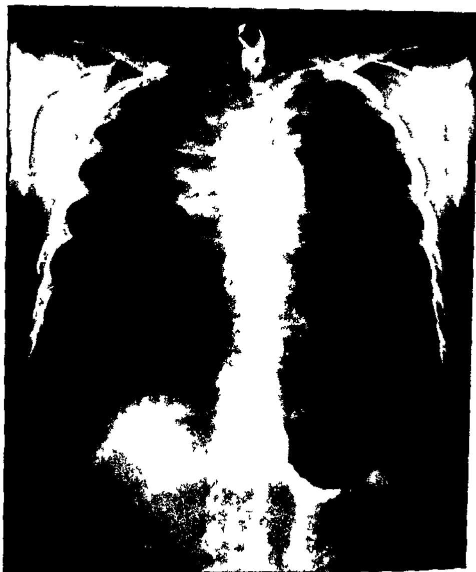


FIGURE 85 A 14 year old boy had a history of swelling of the neck and face of 6 days duration dysphagia fever and dyspnea The esophagogram above was reported as showing a nodular mass in the anterior superior mediastinum presumably malignant Operation was performed by Dr Robert R Linton who removed a malignant thymoma after which the patient did well (Courtesy of Dr John Cass and Dr Robert R Linton)

Lesions of the mediastinum

Intrathoracic goiter In 6 cases of posterior mediastinal goiter Sweet (11) reported only 1 in which the patient had difficulty in swallowing

Mediastinal tumors Benign or malignant mediastinal tumors of such size and location as to press on the esophagus may cause some esophageal obstruction Among these tumors the ones most likely to involve the esophagus are carcinoma sarcoma and lymphoma Benign tumors may cause considerable deviation of the esophagus without significant obstruction

Hill and Vinson (6) studied 50 consecutive cases of mediastinal tumor with reference to pressure on trachea bronchi and esophagus In 32 cases pressure on the air passages was demonstrated with evidence of dyspnea in 31 Pressure on the esophagus was demonstrated in 11 instances but dysphagia was noted in only 4 patients In 3 of the 4 patients who had dysphagia it was mild and was not the major complaint in the fourth patient it was the outstanding complaint Since the esophagus is an elastic mobile tube dysphagia is seldom caused by extrinsic pressure whereas the trachea and bronchi being more rigid are frequently narrowed by external tumors, with resulting dyspnea Of the 4 cases in which dysphagia was present 1 was due to aneurysm of the descending aorta 2 were due to aneurysm of the aortic arch and 1 was due to mediastinal lymphosarcoma

Granulomas In 1955 Garamella Stutzman Varco and Jensen (4) reported 3 uncommon cases of subcarinal mediastinal granulomas that had produced esophageal obstruction They also reviewed 36 reported cases of mediastinal granulomas that had been surgically treated They believe that the probable tuberculous origin of these granulomas demands continued attention

Mediastinal sarcoid also may cause pressure on the esophagus through invasion of mediastinal lymph glands The preoperative x ray study may suggest inoperable malignant neoplasm but exploratory thoracotomy is indicated because it may reveal a

comparatively benign situation such as mediastinal granuloma (sarcoid or tuberculosis)

Tracheobronchial carcinoma may cause extrinsic pressure on the esophagus directly or through metastases to surrounding



Figure 86. Carcinoma of bronchus invading the esophagus, with dysphagia as the first symptom (Courtesy of Dr R H Sweet)

lymph glands Such extrinsic pressure may cause deviation of the esophagus with dysphagia but it is more likely to invade the lumen of the esophagus and produce a fistula (Fig 86)

Dilated left auricle Extrinsic pressure on the esophagus from an enlarged heart is rather unusual (Fig 87) Hurst Messer, and Bland (7) have reviewed their experiences with the enormous hearts seen in rheumatic mitral-valve disease observed at the Mas-



FIGURE 87 X ray showing pressure defect on the esophagus produced by the heart when the patient was in certain positions. There was only slight dysphagia.

sachusetts General Hospital The left atrium contributes greatly to the heart size, but the right atrium and the right and left ventricles are enlarged as well The predominant lesion producing these enormous hearts is rheumatic mitral-valve disease with regurgitation When the heart reaches such a tremendous size



FIGURE 88 Rheumatic heart disease with mitral regurgitation Marked enlargement of the left atrium with narrowing of the barium-filled esophagus. There was some dysphagia (Courtesy of Hurst J Willis Messer Addison L and Bland Edward F Extremes of heart size in mitral valve disease Personal communication)

x-ray examination shows esophageal narrowing and some abnormal deviation of the barium-filled esophagus (Fig 88) Hurst Messer, and Bland (7) described one patient in this group who complained of food sticking in his throat and was careful to chew his food into fine particles for that reason This patient died and autopsy revealed an enormous heart that filled the lower one-third to one-half of each pleural cavity The left atrium extended *to the right chest wall and had a capacity of 2,500 cc* The right and left ventricle were enlarged as well and the right atrium held 250 cc The esophagus the trachea and the right main bronchus were compressed by the gigantic left atrium

Vascular anomalies compressing the trachea or esophagus have been thoroughly discussed by Gross and Neuhauser (5) They include double aortic arch right aortic arch with a left ligamentum arteriosum anomalous innominate artery anomalous left common carotid artery and aberrant subclavian artery Patients with these anomalies should be investigated by fluoroscopic and film studies of the esophagus and by endoscopic visualization of the esophagus and the trachea Surgical treatment is possible in nearly all these types of vascular anomalies In adults and older persons one should search for other causes of dysphagia In 15 cases of dysphagia associated with an aberrant right subclavian artery Resano and Barrani (10) found 4 patients with other conditions causing dysphagia—gastric tumor megaesophagus scleroderma and a short, malformed esophagus

Aortic aneurysm Aneurysm is a possible cause of dysphagia although many aneurysms may press upon the esophagus without producing significant difficulty in swallowing (Fig 89)

Diagnosis

Diagnosis depends on the ability of the physician to ascertain the various causes of pressure on the esophagus It also depends very greatly on the ability of the radiologist to differentiate ex

trinsic pressure on the esophagus and intrinsic disease of the esophagus



FIGURE 89 X ray appearance of a deviated esophagus due to aortic aneurysm The patient was a man 73 years old who had a 6-month history of dysphagia He was doing well 2 months after wiring of the aneurysm

In doubtful cases esophagoscopy should be performed to aid in the differential diagnosis An experienced esophagoscopist can always determine whether or not the disease involves the esophageal mucosa If it does involve the mucosa it must be intrinsic esophageal disease, either primary in the esophagus or actually invading the esophagus from outside If it does not involve the mucosa it must be either a submucosal tumor or an extrinsic mass. Submucosal tumors are usually benign but gastric carcinoma may invade the lower esophagus submucosally

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Treatment

The treatment of masses causing pressure on the esophagus depends on treatment of the underlying disease

Prognosis

The prognosis is that of the underlying disease

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TRAUMA

25

Trauma to the esophagus may result from internal perforation external injury or spontaneous rupture

INTERNAL PERFORATION

Perforations generally occur at sites of normal and pathologic narrowing of the esophagus

Etiology

The esophagoscope the dilating bougie and swallowed foreign bodies in that order are the commonest causes of perforation Statistically the esophagoscope produces perforations at the pharyngoesophageal level the dilating bougie just above the diaphragm and foreign bodies wherever they may lodge

Faulty instrumentation may cause perforation of the esophagus The walls are so thin and delicate especially in elderly women that the greatest care and skill must be used Particular care must be taken when the esophagoscope is at the level of the cricopharyngeus for at this level undue pressure against the unrelaxed muscle may easily tear a hole in the posterior wall of the esophagus where it lies against the bodies of the vertebrae I ld

erly, cachectic, or dehydrated patients are the most vulnerable Instrumental removal of sharp foreign bodies or of foreign bodies which have been in the esophagus for some time, with resulting infection, also is fraught with danger of perforation

Blind bouginage is very likely to cause perforation of the esophagus Except for the blunt mercury-filled rubber covered bougies used in achalasia, bougies should never be passed blindly Fluoroscopy is of no help whatsoever as a guide to bouginage except that in cases of achalasia and megaesophagus it may demonstrate whether the mercury bougie has passed into the stomach or has curled up in the esophagus All other bougies should be passed under direct vision through the esophagoscope or with the use of a previously swallowed thread as a guide

Foreign body Sharp foreign bodies lodged in the esophagus are very likely to perforate if not promptly and carefully removed The perforation may be caused by the sharpness of the foreign body, infection and peristalsis may be contributory factors

Pathology

Simple through-and through perforation with complicating mediastinitis is the usual course of events However, a simple laceration of the mucosa followed by intramural suppuration eventually involving the periesophageal tissues produces the same end result Perforations may result from the pressure necrosis of a retained and impacted foreign body Goligher (12) believes that pressure may occur between the esophagoscope and the protuberance of a hyperextended cervical spine The pinched area could then be involved in a pressure necrosis with symptoms delayed several days until the necrotic tissue had separated and a leaking perforation had occurred

Symptoms

The commonest and earliest symptom of perforation is pain in the neck or chest which is increased by attempts to swallow The

patient may rapidly develop a high fever and progress to a shock-like state, with a thready pulse and hypotension

Diagnosis

Diagnosis is based on a history of pain following instrumentation or ingestion of a foreign body The patient is usually febrile, and his general condition may be deteriorating The blood picture is usually of no help in diagnosis (35) Tenderness and crepitus may be found in the neck, the latter establishes the diagnosis X-rays may reveal widening of the superior mediastinum anterior displacement of the trachea in the neck emphysema in the neck or mediastinum and fluid or air in the pleural space Occasionally Lipiodol swallow may be utilized to confirm the level of perforation (Fig 90) Esophagoscopic examination is not indicated

Frequently the endoscopist will recognize or suspect that a perforation has occurred, but this is not always true Accordingly all patients should be carefully observed during the post-endoscopy period

Perforations of the lower esophagus frequently produce a clinical picture of upper abdominal pain, spasm and tenderness indistinguishable from an acute upper abdominal catastrophe The commonest error in diagnosis is that of a perforated peptic ulcer Numerous regrettable laparotomies have been performed in these circumstances

Treatment

Numerous authors (4, 22 32) have suggested a conservative program nothing by mouth intensive chemotherapy and careful observation for 'small or minor tears particularly if there is no evidence of communication with the pleural cavity Neuhof and Jemerin (25) however found that the mortality in patients treated conservatively before 1936 was 77 3 per cent whereas after that date when nearly all underwent surgery it was



FIGURE 90 Perforation of the lower esophagus caused by a resident who performed bouginage with larger and larger bougies in too rapid succession at the same sitting

17 per cent They did not believe that penicillin and other antibiotics played a significant role in this improvement Weisel and Raine (35) operated on 7 successive patients with traumatic perforations which were from 3 to 38 hours old All were treated by closure of the perforation, drainage, antibiotics, and supportive therapy, and all recovered Overstreet and Ochsner (27) obtained excellent results in 13 cases by following a similar program

We are fully aware that a certain number of these perforations will do very well under conservative management However it is our opinion that a greater over all salvage will be achieved through an aggressive policy of early surgical intervention Cervical mediastinotomy carried out through the standard cervical incision (page 60) will permit evacuation of infected material, drainage, and repair of the esophageal rent with two layers of fine interrupted silk sutures If the tear is in the thoracic esophagus we have utilized a right thoracotomy (page 61) for the upper esophagus and a left thoracotomy (page 63) for the lower esophagus The optimum time for esophageal closure is within the first 6 hours, but successful repairs have been accomplished after as long an interval as 42 hours (27) After closure of the tear the mediastinum and pleura are carefully cleansed of necrotic tissue and foreign matter, the mediastinal pleura left open a chest catheter placed close to (but not on) the esophageal repair and the lung carefully re-expanded We have inserted a second catheter anteriorly to ensure inflation of the lung and minimize the consequences of empyema

Occasionally a patient may be seen several days after perforation has occurred with a well localized abscess in the neck or mediastinum Simple drainage should be promptly instituted In the case of an abscess of the thoracic mediastinum the approach should be retropleural, with a costotransversectomy being used to prevent empyema

Late complications in the form of esophageal fistulas are sometimes seen An esophagocutaneous fistula will almost always heal spontaneously if there is no distal obstruction in the esophagus



FIGURE 90 Perforation of the lower esophagus caused by a resident who performed bouginage with larger and larger bougies in too rapid succession at the same sitting

17 per cent They did not believe that penicillin and other antibiotics played a significant role in this improvement Weisel and Raine (35) operated on 7 successive patients with traumatic perforations which were from 3 to 38 hours old All were treated by closure of the perforation drainage, antibiotics, and supportive therapy, and all recovered Overstreet and Ochsner (27) obtained excellent results in 13 cases by following a similar program

We are fully aware that a certain number of these perforations will do very well under conservative management However it is our opinion that a greater over-all salvage will be achieved through an aggressive policy of early surgical intervention Cervical mediastinotomy carried out through the standard cervical incision (page 60) will permit evacuation of infected material drainage, and repair of the esophageal rent with two layers of fine interrupted silk sutures If the tear is in the thoracic esophagus we have utilized a right thoracotomy (page 61) for the upper esophagus and a left thoracotomy (page 63) for the lower esophagus The optimum time for esophageal closure is within the first 6 hours, but successful repairs have been accomplished after as long an interval as 42 hours (27) After closure of the tear the mediastinum and pleura are carefully cleansed of necrotic tissue and foreign matter, the mediastinal pleura left open a chest catheter placed close to (but not on) the esophageal repair and the lung carefully re expanded We have inserted a second catheter anteriorly to ensure inflation of the lung and minimize the consequences of empyema

Occasionally a patient may be seen several days after perforation has occurred with a well-localized abscess in the neck or mediastinum Simple drainage should be promptly instituted In the case of an abscess of the thoracic mediastinum the approach should be retropleural with a costotransversectomy being used to prevent empyema

Late complications in the form of esophageal fistulas are sometimes seen An esophagocutaneous fistula will almost always heal spontaneously if there is no distal obstruction in the esophagus



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FIGURE 91 Traumatic diverticulum of the esophagus caused by shrapnel. The diagnosis was made by x ray examination and confirmed by esophagoscopy Relief was obtained by radical surgery

and no foreign body in the fistulous tract An esophagotracheal fistula will require surgical repair Persistent empyema fed by an esophageal sinus will require rib resection and drainage An esophagopulmonary fistula is a serious complication, once the location of the fistula is established (by bronchoscopy, bronchography, esophagoscopy, and Lipiodol swallow as necessary), thoracotomy with repair of the esophagus lobectomy, and drainage will be necessary If traumatic perforation is treated conservatively a traumatic diverticulum may develop which may later require surgical repair (Fig 91)

Prognosis

Assuming that the patient is in good general condition, prompt recognition and repair, with proper drainage and utilization of antibiotics, usually provides an excellent result The prognosis becomes progressively poorer with delay in diagnosis and treatment be it expectant or surgical

External Injury

Etiology

External injury to the esophagus may be caused by bullets shrapnel, glass or other sharp objects Gunshot or stab wounds of the thorax are usually complicated by perforation (Fig 92)

Diagnosis

Unfortunately an esophageal wound is frequently overlooked in the care of the patient with a perforating thoracic or cervical wound The history of the accident dysphagia spitting up of blood supraclavicular emphysema and pain on swallowing should indicate the diagnosis X ray findings are similar to those in internal perforation



FIGURE 92 A 45 caliber bullet shattered the patient's right mandible traversed the neck in a downward direction and lodged in the right supraclavicular space Air can be seen in the retropharyngeal space The patient did well after removal of the missile and drainage of the superior mediastinum

Treatment

The treatment is prompt surgery as described in the previous section, in addition to the repair of any surrounding structures which also may have been damaged

Esophagoscopy and bouginage may be indicated in occasional cases to dilate a post traumatic stricture

SPONTANEOUS RUPTURE

Spontaneous rupture of the esophagus represents a traumatic perforating tear of the esophagus almost always in its lower third, probably due to sudden increases of intraluminal pressure The term "spontaneous rupture is both contradictory and inaccurate and the suggestion has been made that the term pressure perforation (23) or "postemetic rupture (31) be substituted Spontaneous rupture has however acquired the dignity of usage and acceptance and we shall accordingly refer to it as such

Etiology and pathology

Rupture in this condition is believed to represent a blowout of the lower esophagus secondary to a sudden increase of intra-abdominal pressure Such tears are most commonly associated with vomiting but have been reported after defecation (36) and during weight lifting (13) seizures (16) labor and abdominal trauma (14 24 28) This sudden rise in intra abdominal pressure forces gastric contents abruptly into the esophagus by compressing the stomach (Fig 93) The sudden rise in intraluminal pressure coupled with a closed cricopharyngeal pinchcock ruptures the esophageal wall in an explosive fashion Although a pre-existing esophagitis is believed by some (6 1) to be necessary before such perforation occurs most reported cases have occurred in normal esophagi Doubtless other factors such as anatomic or acquired weakness of the wall neuromuscular incoordination chemical damage to the mucosa and general debility contribute to the actual occurrence (5)

Samson (31) believes that contraction of the spiral musculature of Lerche may play an important role in rupture The experiments of Mackenzie (18) and Mackler (19) on cadavers suggest that such a special mechanism is not necessary In their experiments esophagi were inflated under pressure until rupture occurred All tears were longitudinal and were located in the lower esophagus,

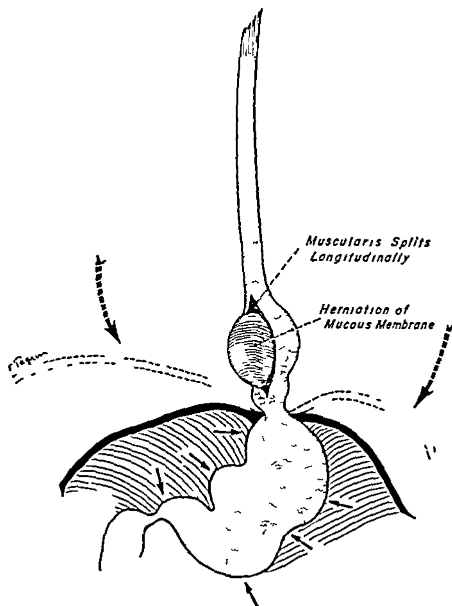
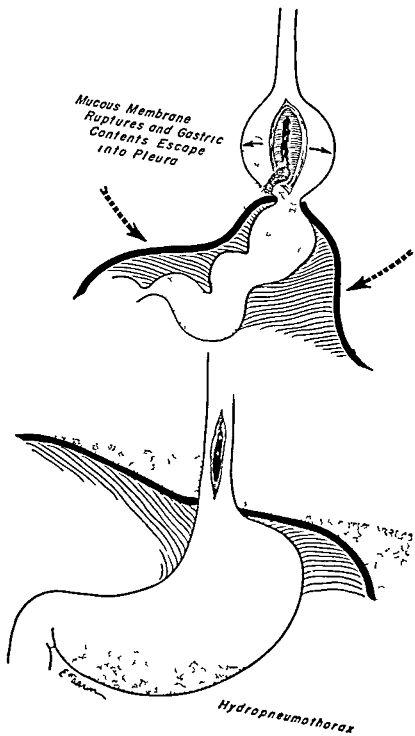


FIGURE 93 Mechanics of spontaneous rupture A sudden rise of intra abdominal pressure coupled with the descent of the diaphragm forces gastric contents into the esophagus With a closed glottis the sudden rise of intraluminal pressure results in perforation with escape of gastric contents and air into the thorax


Mucous Membrane
Ruptures and Gastric
Contents Escape
into Pleura
Hydropneumothorax

the mucous membrane was the last coat to burst The muscularis was first observed to split longitudinally, after which a herniation of mucous membrane ballooned out through the split and rupture followed

Mallory and Weiss (20) described longitudinal mucosal lacerations in the cardia of the stomach at autopsy in patients with exsanguinating hemorrhage They considered that the preceding retching and vomiting were the main causative factors It is likely that the syndrome represents an incomplete rupture of the esophageal *wall extending through the mucosa and submucosa which* occurs in the thoracic portion of a stomach involved in a hiatus hernia (10)

Symptoms

An apparently healthy person suddenly begins to vomit after gastronomic excess and rapidly develops excruciating epigastric pain extending through the left side of the chest to the back, *followed by collapse shock dyspnea, and cyanosis The vomiting* may continue but usually ceases and the patient develops marked thirst The pain is intense and persistent and is not lessened by large doses of morphine Occasionally there is no history of a preceding large meal the vomiting being due to a neurogenic lesion (17) postoperative nausea or some other condition Sometimes there is no true vomiting merely severe and repeated retching or rarer still there is no vomiting or retching whatsoever, rupture occuring apparently as a primary condition (29) The *stomach or duodenum (2) A small number have a history of mild* brown coffee-ground material, occasionally associated with a small amount of blood and more rarely with free bleeding

Diagnosis

A few patients present a history of previous ulcer of the *material ejected during vomiting or retching may be food or dark*

dysphagia and substernal distress suggestive of esophagitis or esophageal stricture, with impaction of a bolus of food at the stricture site, the rupture taking place during a strenuous attempt to force the food past the narrowed area The interested reader should not fail to read Barrett s excellent review (2)

Kinsella *et al* (15) state that the clinical picture of spontaneous rupture of the esophagus is not generally recognized even though its symptoms are generally quite characteristic Rupture usually occurs during a bout of vomiting In 66 per cent of cases there is cervical emphysema Frequently there is rigidity and spasm of the upper abdomen which may result in a misdiagnosis of perforated peptic ulcer (8 15) Mackler (19) considers the triad of vomiting low thoracic pain and emphysema to be diagnostic Physical examination of the chest may be reported as negative, but frequently a pneumothorax with fluid on one or both sides — more frequently on the left — may be found increasing as the period following perforation lengthens The condition must be differentiated from acute perforation of a peptic ulcer acute pancreatitis spontaneous pneumothorax acute coronary occlusion cholecystitis mesenteric thrombosis and dissecting aneurysm Usually there is little difficulty in making the diagnosis if one thinks of it X ray examination may demonstrate mediastinal emphysema and a hydropneumothorax A Lipiodol (not barium) swallow may demonstrate the perforation The presence of gastric contents in aspirated pleural fluid is pathognomonic

This emergency is seen almost exclusively in men It is probably not a sex predisposition but more likely related as Barrett (2) suggests to a male inclination to indulge in pursuits which predispose to vomiting

Treatment

The only treatment is immediate surgery with supportive measures

In this condition the tear is almost always located in the left

posterolateral aspect of the lower portion of the esophagus Only rarely has spontaneous rupture of the esophagus been reported (9, 30) in the upper portion of the gullet Accordingly, the standard left thoracotomy approach is almost always indicated Barrett (3) in 1947 was the first successfully to treat a patient with spontaneous perforation In the same year Olsen and Clagett (6) reported a successful repair The technical maneuvers are identical to those for a traumatic perforation of the lower esophagus (3) If there is any unavoidable delay in closure of the esophageal tear, the pleural space should be drained of fluid and air by closed thoracotomy and the patient should be given fluid and blood intravenously and antibiotics parenterally, the latter should be administered in large doses We have not utilized local chemotherapy at the time of thoracotomy Intensive administration of systemic antibiotics is, of course, invaluable A nasogastric tube may be passed in the course of the repair to aid in gastric evacuation and to keep the stomach deflated after operation A feeding jejunostomy may be a lifesaving complementary procedure in the debilitated patient

Prognosis

In 1951 only 13 of the approximately 100 patients with spontaneous rupture reported in the world literature had survived (11) By 1953 23 successful cases had been reported (34) Since then many additional cases have been recorded (1, 7, 8, 33) This high mortality rate is caused by the virulent mediastinitis and pleuritis that are associated with cardiorespiratory difficulties secondary to mechanical compression However even though the patient's condition be critical, if surgical repair and drainage can be instituted within a few hours the prognosis is usually good The two chief factors in a successful outcome are establishing the diagnosis and instituting prompt surgical treatment The longer this combination is delayed, the worse the prognosis

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Esophageal Atresia and Fistula

26

Esophageal atresia represents a congenital lack of continuity of the lumen of the esophagus there may be complete absence of a long segment of esophagus or there may be only a short narrow imperforate segment

Bronchoesophageal and tracheoesophageal fistulas are abnormal communications between bronchus and esophagus or between trachea and esophagus These may be congenital or acquired

Congenital Defects

By the eighth week of intrauterine life the trachea and esophagus should be completely separated and normally developed (see Chapter 2) Any developmental anomalies have been established by this time The cause of anomalies such as persistence of a communication between the two (fistula) or obliteration of the esophageal lumen (atresia) is unknown It is generally believed that the two lateral septa of the cephalic extremity of the foregut fail to meet and fuse as they should in order to differentiate the upper half of

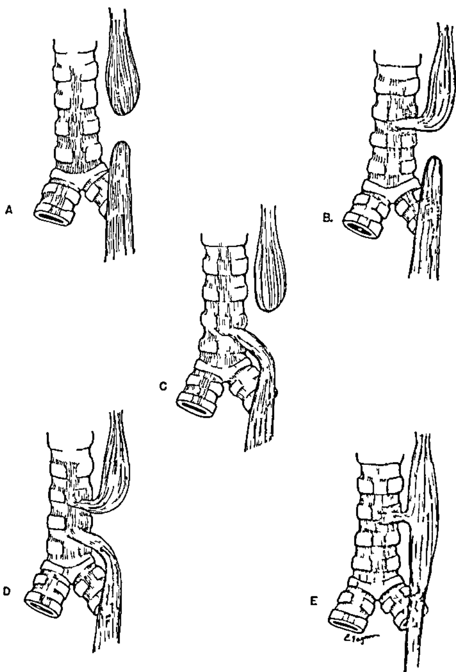


FIGURE 94 Types of congenital tracheoesophageal fistula (modified from Gross)

the esophagus from the trachea The embryologic aspects of such malformations are well reviewed by Ferguson (8) and Haight (11)

The arrangement of the trachea and the esophagus in the various types of esophageal atresia and tracheoesophageal fistula has been well shown diagrammatically by Gross (Fig 94) The upper segment of the esophagus may end blindly (Fig 94A) or may communicate with the trachea (Fig 94B) The lower segment of the esophagus may start from a blind end (Fig 94A and B) or may start from a communication with the trachea (Fig 94C) Still another arrangement is that in which both segments of the esophagus communicate with the trachea but not with each other (Fig 94D) Finally, a fistula may exist without esophageal atresia (Fig 94E)

Esophageal atresia with tracheoesophageal fistula

This anomaly occurs approximately once in every 2000 births A fistula is to be found in at least 80 per cent of patients with congenital atresia, type C of Figure 94 wherein the communication occurs between the lower part of the esophagus and the trachea, is by far the most common

Diagnosis

A newborn infant that shows difficulty in breathing with excessive mucus and saliva in mouth and pharynx should be immediately suspected of this anomaly Choking and cyanosis may occur at feeding There may be a history of immediate vomiting following the first and each successive feeding Acute abdominal distention may be noted, this results from the pumping of air into the gastrointestinal tract through the fistula

A soft red rubber (radio-opaque) catheter is passed down the esophagus where an obstruction is promptly met Instillation of a small amount of contrast medium (never barium) will demonstrate the presence of a fistula (Fig 95) A lateral view will provide the best visualization The latter is suspect by pressure of gas in the intestines in the plain film (Fig 96) The latter may also be sug

gestive of associated anomalies such as duodenal or jejunal atresia or imperforate anus Endoscopy has not been utilized as a diagnostic aid in these cases

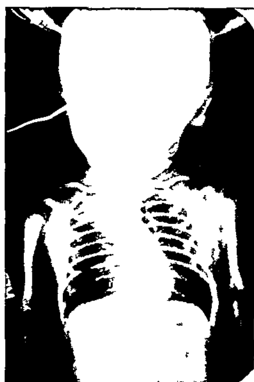


FIGURE 95 Instillation of excessive contrast medium results in overflow and pulmonary aspiration

Associated anomalies

A very large number of these patients have associated congenital anomalies In Haight s (11) series 58 per cent had associated anomalies of life threatening significance In order of frequency they were congenital heart disease and gastrointestinal neurological, and genitourinary disorders The gastrointestinal anomalies such as duodenal obstruction or imperforate anus are of particular importance since they are frequently remediable and demand immediate correction

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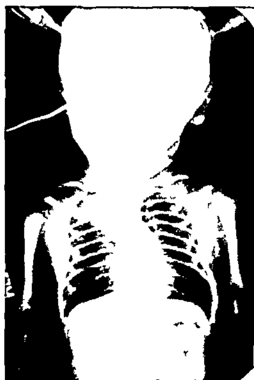


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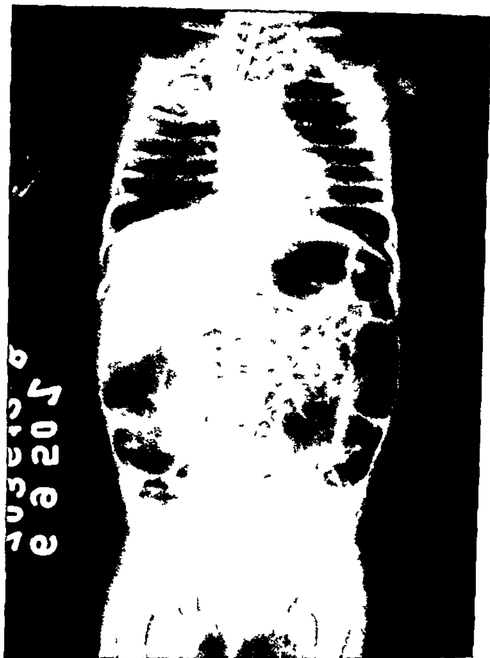


FIGURE 96 X ray of baby girl one day old showing correct technique in the diagnosis of congenital atresia This x ray was reported as follows The lungs are bright except for an area of increased density in the right upper lobe A catheter extends down the esophagus to the level of the third thoracic vertebra Just below the tip of the catheter the esophagus appears to stop abruptly and above it is rather markedly dilated and filled with air A large amount of air is seen within the intestinal tract down to the region of the sigmoid Findings are consistent with atelecta sis or aspiration pneumonitis of the right upper lobe atresia of the esoph agus and imperforate anus or distal rectum

Treatment

Prompt and effective surgery is the infant's only chance for survival. The first successful primary anastomosis of the esophagus for esophageal atresia was performed by Haight in 1941 (12). Surgery is never undertaken as an emergency procedure, and proper steps should be taken to provide adequate hydration, chemotherapy, and clinical evaluation. Special nursing care to keep the pharynx clear of secretions with constant suction should be provided. The infant should be placed in a humidified oxygen tent. A saphenous vein cutdown is inserted and fluids, blood, and chemotherapy administered parenterally with the cooperation of a pediatrician. During this period the infant is seen by an anesthetist experienced in pediatric surgery and plans made for operation. These infants are poikilothermic, and their temperature should be watched carefully before, during, and after operation.

We have preferred a right transthoracic approach through the fourth or fifth intercostal space. The azygos vein is divided and the upper esophageal segment is dissected free well up into the neck (Fig. 97A). It is held by transfixion stitches rather than clamps. Next the lower esophageal segment is mobilized. The vagi are identified and carefully spared. Gross (10) has advised starting this dissection at the mid-portion of the lower segment and working upwards, thus minimizing the chance of tearing the left pleura and hence adding to the anesthetist's difficulties and the patient's morbidity. The dissection is carried down only enough to allow the anastomosis to be completed without tension, for unlike that of the upper segment, maintenance of the blood supply at this level may be vitally important.

The fistula is then carefully divided flush with the trachea and the tracheal defect is carefully sutured with interrupted stitches of 5-0 silk. If possible, a flap of pleura is placed over this closure.

The thin and atretic lower esophagus is gently dilated and a full thickness button excised from the upper pouch (Fig. 97 B, C and D). A two layer open anastomosis is carried out with interrupted

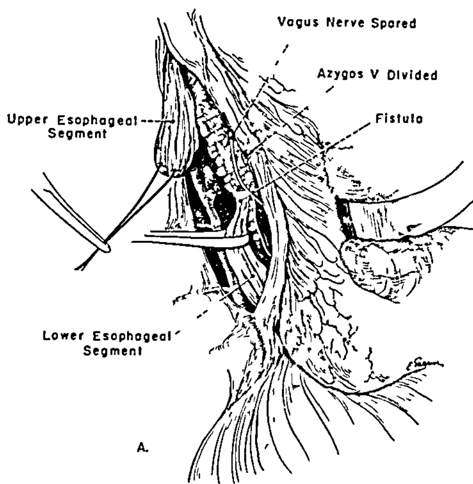


FIGURE 97 Details of dissection in carrying out a primary anastomosis. In (A) note the proximal pouch held by sutures and the technic of dissection of the lower segment The rest of the drawings (B) (C) and (D) show anastomosis of the dilated proximal pouch to the smaller distal segment

5-0 silk sutures Trimming the distal esophagus at an angle will provide a larger lumen Clamps are never utilized for the anastomosis By placing a posterior row of sutures before trimming the distal end the anastomosis will be facilitated and the fragile distal cuff can be sutured with less trauma The mediastinal pleura is loosely re-approximated The lung is expanded a posterior catheter inserted and the chest closed A complementary Stamm gastrostomy is performed at the end of the primary procedure This may

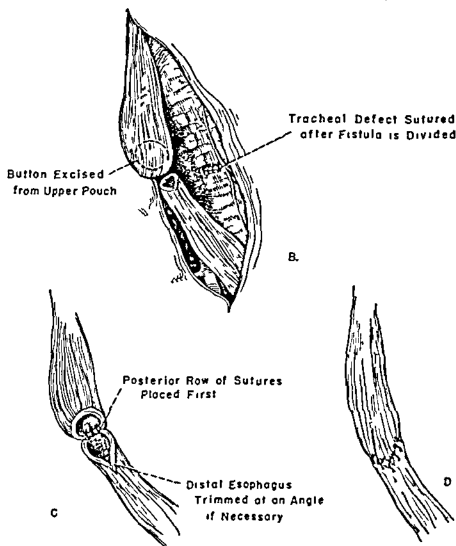


FIGURE 97 (*continued*) See legend page 326

be deferred for 48 hours Though not always necessary it is usually advisable During this procedure care should be taken to anchor the gastric wall to the parietes Gastrostomy feedings are continued until the infant can maintain itself satisfactorily — a period of about 10 days When no longer necessary the rubber feeding tube is removed and the gastric fistula closes spontaneously and uneventfully

Occasionally the two esophageal *segments cannot safely be* brought together In these cases the fistula is taken down and the tracheal defect closed as described The distal segment of the esophagus is closed and the thoracic portion of the procedure terminated An incision is made in the neck along the left sternomastoid (Fig 98), the esophagus mobilized, and the proximal segment drawn out and the pouch opened and sutured to the skin A complementary gastrostomy is then carried out

Restoration of the gastrointestinal continuity is effected 18 to *24 months later This may be carried out by mobilizing either the* stomach or the colon To mobilize the stomach the left thorax is entered through the eighth intercostal space and the diaphragm divided (Gross [10] has been able to mobilize the stomach by dissecting the hiatus and dividing the left gastric artery without opening the diaphragm) The distal portion of the esophagus is excised, the cardia and gastrostomy stoma are closed, and the stomach mobilized as described in Chapter 19 for a supra aortic anastomosis The diaphragm is repaired, the lung expanded, and the chest closed with drainage

An incision is then made over the cervical stoma the superior *segment is mobilized, and a cer*vical esophagogastrostomy is accomplished with 2 layers of interrupted silk sutures (Chapter 19) The neck wound is closed without drainage and the cutaneous gastrostomy stoma repaired

We are inclined to believe that as more experience with substernal colon transplant is gained this method will be preferred to esophagogastrostomy as a delayed procedure in this condition There are numerous reports *of successful cases at the present time* (5, 11 21, 2.) The technic is identical to that described in Chapter 12, page 132

Complications

The commonest complication and the most frequent cause of death *is the combination of atelectasis* and pneumonia Technical

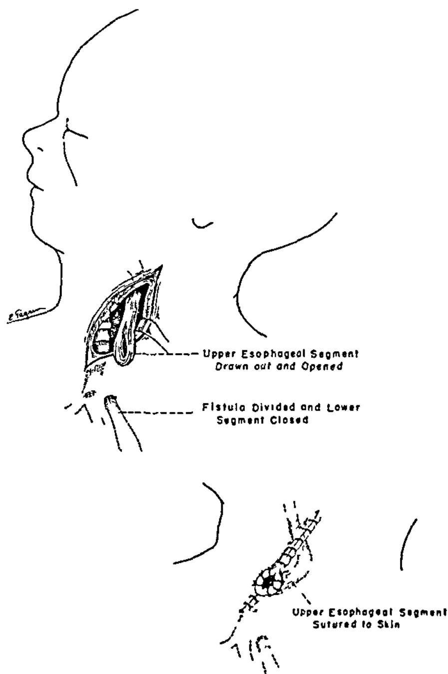


FIGURE 98 Creation of a cervical esophageal fistula where repair is to be staged

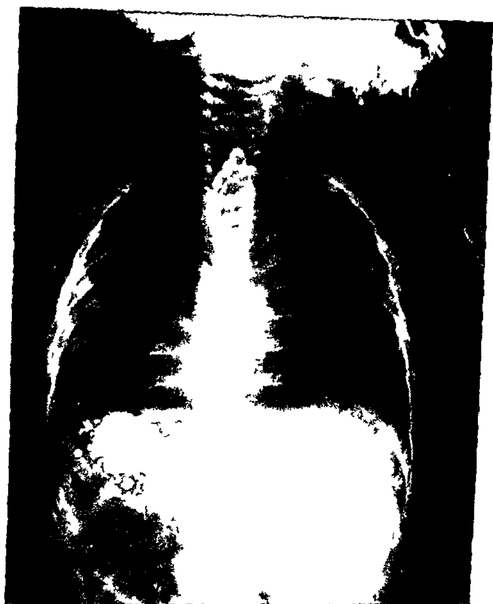


FIGURE 99 *Congenital atresia of the esophagus 11 months after surgical repair* Almost complete esophageal obstruction existed due to foreign bodies in the esophagus These foreign bodies consisted of multiple wads of paper and cotton with fragments of oilcloth from toys (See Figure 100) Gastrostomy *had been advised in another clinic but* endoscopic removal of the foreign bodies and bouginage gave relief

complications associated with surgery such as leakage or disruption of the anastomosis or recurrence of the tracheoesophageal fistula are not uncommon Esophagocutaneous fistulas will heal spontaneously with proper drainage Late complications may occur with stenosis at the suture line, this may in turn lead to aspiration or perforation from bouginage In general bouginage is effective in dilating postoperative strictures (Figs 99 100) When such strictures are severe or recalcitrant to dilatation reoperation is probably the safest course Of 97 living patients with esophageal anastomosis reported by Gross (10) 34 required dilatation and 4 reoperation

Cough is frequently a disturbing postoperative complication it is usually caused by the sutures in the trachea A persistent cough, however suggests a recurrent fistula Such recurrences are best demonstrated roentgenologically using iodized contrast media with the baby in a prone position (26) Although these occasionally may close spontaneously reoperation is safer and more effective

Prognosis

Mortality and morbidity vary with prematurity type of anomaly, general condition of the infant when first seen and the presence and type of associated anomalies (9) If the case is not treated death from aspiration pneumonia usually occurs within a week

The over all mortality rate in a large series varies between 40 and 80 per cent The surgical mortality lies between 20 and 50 per cent

Roberts *et al* (23) have subdivided Gross s type C (see Figure 94) into subgroups C1 and C2 (Fig 101) In their experience the first type, with a short upper end are unfavorable for primary anastomosis and the C2 group with overlapping esophageal ends are most favorable for primary anastomosis and offer an excellent prognosis



FIGURE 100 Same case as Figure 99 showing esophageal obstruction relieved after esophagoscopic foreign body removal

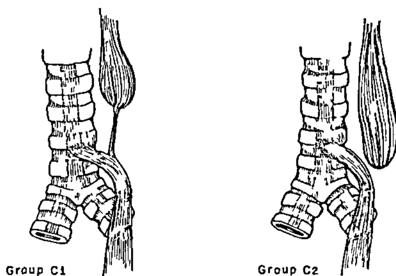


FIGURE 101 Roberts's sub division of Gross's type C (see Figure 94) into C1 and C2 The presence of a long upper pouch is most favorable for primary anastomosis

Esophageal atresia without fistula

This anomaly occurs in approximately 10 per cent of infants with esophageal atresia The diagnostic clue of absence of a fistula is in the roentgenogram which fails to reveal the presence of air in the stomach or intestinal tract At the time of surgery one usually finds a partial agenesis of the esophagus with only a stump extending 2 or 3 cm above the diaphragm This rudimentary lower esophagus does not reach high enough to communicate with the trachea Such patients require mobilization of the stomach into the apex of the chest at the time of the initial procedure or a cervical esophagostomy and feeding gastrostomy When the child has reached 18 to 24 months of age, an esophagogastrostomy or colon substitution can usually be accomplished safely and with ease In general we prefer the latter procedure

Because of its rarity, statistical results in this group are not reliable In general the prognosis is poor Battersby (2) had not a single survival in 6 such cases

Congenital fistula without atresia

Lamb, (15) in 1873, was the first to report a case of tracheo-esophageal fistula without other esophageal abnormalities In 1954 Ware and Cross (30) summarized 27 cases of this anomaly to which *Haight (11) added 7, making a total of 34* The condition has been admirably reviewed by Ferguson (8)

The diagnosis is difficult, since the condition *is rare and is infrequently* considered The clinical features are paroxysms of coughing at feeding, gaseous distention of the gastrointestinal tract and persistent pneumonitis Sieber and Girdany (27) have passed a tube into the stomach and observed continued filling of the stomach with air under pressure when the infant cries or inspires vigorously In this condition endoscopy may be a very valuable procedure, and it *should be performed whenever the location of the* fistula is in doubt The fistula can usually be demonstrated by instilling an iodized preparation through a catheter in the esophagus (13) Sarkissian (24) found a congenital fistula 0 5 cm below the level of the cricoid cartilage, repair was accomplished through the neck

These patients should have immediate repair, they have the most *favorable prognosis of any in this group (2, 28)*

ACQUIRED FISTULA

Etiology

Acquired fistulas between the esophagus and trachea or bronchus may be due to any one of the following conditions (arranged in order of frequency)

(1) Neoplasm (17)
 (a) Carcinoma (Fig 102)
 (b) *Hodgkin s disease*
 (c) Sarcoma *(25)*

(2) *Infections*
 (a) Tuberculosis (6 20)



Figure 102 X ray appearance of carcinoma of the esophagus with esophagobronchial fistula The x ray shows a large ulcerating tumor extending from the midpoint to the cardia with barium flowing through a small fistula into the right lower lobe bronchi

Congenital fistula without atresia

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The diagnosis is difficult, since the condition is rare and is infrequently considered The clinical features are paroxysms of coughing at feeding, gaseous distention of the gastrointestinal tract and persistent pneumonitis Sieber and Girdany (.7) have passed a tube into the stomach and observed continued filling of the stomach with air under pressure when the infant cries or inspires vigorously In this condition endoscopy may be a very valuable procedure, and it should be performed whenever the location of the fistula is in doubt The fistula can usually be demonstrated by instilling an iodized preparation through a catheter in the esophagus (13) Sarkissian (24) found a congenital fistula 0 5 cm below the level of the cricoid cartilage, repair was accomplished through the neck

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Acquired Fistula

Etiology

Acquired fistulas between the esophagus and trachea or bronchus may be due to any one of the following conditions (arranged in order of frequency)

(1) Neoplasm (17)
- (a) Carcinoma (Fig 10.)
- (b) Hodgkin s disease
- (c) Sarcoma (25)

(.) Infections
- (a) Tuberculosis (6 .0)

fistulous opening Some cases are demonstrable only by endoscopy, bronchoscopy being preferable to esophagoscopy since the fistulous opening into the esophagus may be hidden from view in a tumor mass or in mucosal folds

Treatment

Treatment will depend on the underlying cause In cases of syphilis, tuberculosis or other infectious disease appropriate specific therapy and antibiotics should be utilized Clerf Cooley, and O Keefe (4) reported two patients with esophagobronchial fistula of unknown etiology in which prompt closure of the esophageal end of the fistula resulted after sodium hydoxide crystals fused on a curved metal applicator were applied esophagoscopically and carried into the fistula for a distance of several centimeters Both *patients were free from symptoms for over eighteen months* We have attempted this procedure without success

In general surgical closure of the fistula is the most effective and satisfactory form of treatment It is preceded by a trial of antibiotic management in cases that are due to syphilis or tuberculosis Surgical repair should be performed in any fistula not due to neoplasm The surgeon should be prepared to carry out a complementary lobectomy or pneumonectomy if pulmonary tissue is involved in the basic process When the fistula is due to malignant disease it is unlikely that surgery will have much to offer However if the patient has a chance of survival closure of the fistula may be attempted for palliative reasons We have recently operated on a young man with a tracheoesophageal fistula secondary to mediastinal Hodgkin s disease and radiation damage The fistula was closed and the right colon was utilized as an esophageal substitute

We prefer a transthoracic approach usually on the right Should any pulmonary involvement be present the incision is chosen accordingly After incision of the mediastinal pleura the esophagus is dissected free above and below the fistula The fistulous tract is

(b) Syphilis (16, 19)
(c) Nonspecific infection (14)
(d) Actinomycosis (29)
(e) Esophagitis
(f) Empyema (7)

(3) Traumatic
(a) Foreign body
(b) Instrumentation
(c) Crushing injury

(4) Diverticula (18)

Incidence

In a study of all types of fistulas between the esophagus and bronchus or trachea, Monserrat (20) found 367 fistulas of neoplastic origin, 222 of congenital origin 41 of infectious origin and 40 of traumatic origin Abbott (1) believes that the most common infectious basis is tuberculosis According to Clagett, Payne and Moersch (3) carcinoma of the esophagus is the most frequent cause of fistula formation Carcinoma of the bronchus mediastinal glands and thyroid gland also may cause esophagobronchial or esophagotracheal fistula One case reported by Abbott (1) was due to Hodgkin s disease The spontaneous formation of a fistula between the esophagus and trachea or bronchus is so common in any carcinoma of the upper or middle portion of the esophagus that all such patients should undergo bronchoscopy before surgical resection is considered

Diagnosis

A very tiny fistula between the esophagus and trachea or bronchus may cause no symptoms whatsoever Often however, there may be choking or strangling when the patient swallows liquids Solid food may be swallowed without any difficulty Careful fluoroscopy with barium or iodized oil may demonstrate the

fistulous opening Some cases are demonstrable only by endoscopy, bronchoscopy being preferable to esophagoscopy since the fistulous opening into the esophagus may be hidden from view in a tumor mass or in mucosal folds

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We prefer a transthoracic approach usually on the right Should any pulmonary involvement be present the incision is chosen accordingly After incision of the mediastinal pleura the esophagus is dissected free above and below the fistula The fistulous tract is

divided flush with the posterior surface of the trachea or bronchus After debridement of the edges closure is accomplished with interrupted sutures of 5-0 silk Only a single layer is utilized for the tracheobronchial repair, the suture line is reinforced with a flap from pleura, lung or adjacent fat pad The esophagus is closed with two layers of sutures one in the mucosa and the other in the muscularis, an attempt is made to reinforce this suture line whenever possible The mediastinal pleura is left open and the chest closed with posterior pleural drainage When such a fistula is located in the neck the entire repair may be carried out through the standard left cervical incision

Prognosis

The prognosis in acquired fistula depends upon the etiology In malignant disease surgery rarely offers more than palliation of repeated bouts of aspiration and pneumonia In nonmalignant disease the prognosis is generally good

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Specific Infectious Diseases

27

Tuberculosis

Tuberculosis is an infection caused by the tubercle bacillus its lesions are characterized by diffuse infiltration tubercle formation, caseation, necrosis, ulceration or fibrosis

Incidence

Tuberculosis of the esophagus is a very rare disease Lockard (21) found only 15 cases of tuberculosis of the esophagus in a series of 16,489 autopsies of tuberculous subjects According to Hamberger (1[illegible]) up to 19[illegible]6 only 116 cases of esophageal tuberculosis had been reported Four cases of esophageal tuberculosis were diagnosed by Pulver and Michel ([illegible]8) from 1949 to 1953 Wexels (40) stated in 1954 that about 1[illegible]5 cases had been reported Most of these cases were first diagnosed at post mortem examination in patients with tuberculosis in other organs especially in the lung In a 10 year period at the Bellevue Hospital New York City Carr and Spain (5) found only 1 case of esophageal tuberculosis in a

series of 1400 autopsies on tuberculous patients The mechanism of infection in their case was implantation of the bacilli in a carcinomatous ulceration of the esophagus

Etiology

In 1913 Lockard (21) presented a comprehensive review of the literature, he classified tuberculosis of the esophagus into the following groups on the basis of their pathogenesis

(1) Direct implantation of bacilli following trauma to the esophagus

(2) Direct implantation of bacilli in a carcinomatous ulceration of the esophagus

(3) Direct implantation of bacilli without a previous lesion of the esophagus

(4) Extension from the larynx or the pharynx

(5) Extension from vertebral caries (36)

(6) Extension from caseous mediastinal lymph nodes

(7) Hematogenous infection

(8) Unclassified

Considering the fact that patients with pulmonary tuberculosis are habitually swallowing infectious material the low incidence of esophageal tuberculosis can probably be accounted for by the protective mechanism of the squamous-cell lining of the esophagus and its smooth tubular structure without crypts or pockets in which exudate might stagnate Furthermore, the esophagus is frequently flushed out by food, drink and saliva, and this prevents prolonged contact of infectious material with the mucosa Lockard believes that extension by continuity from caseous bronchial glands is by far the most common etiologic factor

Pathology

The upper third of the esophagus is most frequently involved The tuberculous lesion may appear as a superficial mucosal ulceration or, more rarely, as a tumor like growth Some of these

lesions progress to sclerosis and progressive stenosis The ulcerative form produces painful deglutition Perforation may occur DeFazio (9) has reported an extremely rare esophagocutaneous fistula of tuberculous origin According to Wexels (40) esophageal tuberculosis may occur in the course of hematogenous spread in miliary tuberculosis

Symptoms

Sometimes there are no symptoms but in some patients dysphagia and substernal pain are prominent Ulcerative lesions are likely to be characterized by odynphagia stenotic lesions result in dysphagia Cough is commonly present but it may be due to associated laryngeal or pulmonary tuberculosis or to the presence of an esophagobronchial fistula

Diagnosis

When a patient with tuberculosis of the lungs larynx trachea, bronchus mediastinum or vertebrae complains of painful or difficult swallowing tuberculosis of the esophagus should be suspected X-ray examination may reveal an obstructing lesion but cannot establish a specific diagnosis

Esophagoscopy Esophagoscopy with biopsy is the only means of making a positive diagnosis Grossly the esophagus may show esophagitis ulceration tubercle formation proliferation thickening scarring or stricture

Treatment

Streptomycin with para aminosalicylic acid has been shown to be beneficial in the treatment of esophageal tuberculosis Newer drugs such as isonicotinic acid hydrazide may prove even more effective In Wexel s proven case of tuberculosis of the esophagus successful treatment was carried out with streptomycin and esophagoscopic bouginage

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The upper third of the esophagus is most frequently involved The tuberculous lesion may appear as a superficial mucosal ulceration or, more rarely as a tumor like growth Some of these

lesions progress to sclerosis and progressive stenosis. The ulcerative form produces painful deglutition. Perforation may occur. DeFazio (9) has reported an extremely rare esophagocutaneous fistula of tuberculous origin. According to Wevels (40) esophageal tuberculosis may occur in the course of hematogenous spread in miliary tuberculosis.

Symptoms

Sometimes there are no symptoms, but in some patients dysphagia and substernal pain are prominent. Ulcerative lesions are likely to be characterized by odynphagia; stenotic lesions result in dysphagia. Cough is commonly present but it may be due to associated laryngeal or pulmonary tuberculosis or to the presence of an esophagobronchial fistula.

Diagnosis

When a patient with tuberculosis of the lungs, larynx, trachea, bronchus, mediastinum, or vertebrae complains of painful or difficult swallowing, tuberculosis of the esophagus should be suspected. X-ray examination may reveal an obstructing lesion but cannot establish a specific diagnosis.

Esophagoscopy. Esophagoscopy with biopsy is the only means of making a positive diagnosis. Grossly the esophagus may show esophagitis, ulceration, tubercle formation, proliferation, thickening, scarring, or stricture.

Treatment

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Prognosis

Before the days of streptomycin the prognosis was considered bad, with esophageal perforation, mediastinitis, and death occurring in some cases. With the advent of streptomycin, however, the prognosis of esophageal tuberculosis should be good, and the general prognosis depends on the extent of the associated pulmonary or other tuberculous lesions.

SYPHILIS

Syphilis is caused by *Treponema pallidum*, which is a spiral curved organism that shows active movement in fresh specimens. Syphilis is a specific disease of slow evolution, and is either acquired or congenital. Acquired syphilis is propagated by inoculation, congenital syphilis is transmitted through the mother. Syphilis may coexist with carcinoma and tuberculosis, as reported by Dean and Gregg (8).

Incidence

Syphilis of the esophagus is exceedingly rare. It is said to occur occasionally in the congenital form. In the primary stage of acquired syphilis chancre could conceivably develop in the mucous membrane of the esophagus, but such reports have not been discovered. In the secondary stage syphilis may give rise to esophagitis. Tertiary syphilis or gumma of the esophagus has been reported by many observers. Three cases of gumma of the diaphragm producing esophageal obstruction have been reported by Kampmeier and Jones (16). In 1950 Hudson and Head (14) reported a case of tertiary syphilis of the esophagus and reviewed the literature. Their case is the only recent and really convincing one reported. It concerned a patient of 56 years with dysphagia, highly positive Kahn and Wasserman reactions, an almost completely occluding spherical tumor at the cardiac end of the esopha

gus seen at x-ray and a flesh-colored finely irregular non ulcerative mass filling the lower esophagus seen at esophagoscopy Biopsy revealed only stratified squamous epithelium Antisyphilitic therapy was given and ' the result was astounding There was total relief of all symptoms and a negative x ray examination within two weeks, sixty days later esophagoscopy was negative Other reports include those of Wile (41) (1914) who gives a good early bibliography, Abel (1) (1928) Avery (2) (1936) and Peterson (27) (1942)

Pathology

Syphilitic lesions of the esophagus do not differ from syphilitic lesions elsewhere in the body Gummas usually arise in the upper third of the esophagus, and when rupture of the gumma occurs a typical gummatous ulcer is produced which is very likely to progress to stricture formation Tracheoesophageal fistula may be of syphilitic origin (4) Aneurysm may cause pressure on the esophagus but seldom produces enough pressure to cause esophageal symptoms Berman (3) has reported an interesting case of syphilitic aortitis with esophageal perforation In that case a saccular enlargement of the distal portion of the arch of the aorta at the level of the fifth thoracic vertebra was displacing the esophagus laterally and slightly posteriorly to the right for approximately 2 5 cm beyond its normal position The esophagus was adherent to this saccular aneurysm and when both structures were opened (at autopsy) there was exposed a perforation extending from the aorta into the esophagus and measuring 8 mm in diameter It was through this opening that the patient had spilled over 2000 cc of blood which was found in the stomach

Symptoms

Dysphagia is likely to be the only symptom of esophageal syphilis However any of the symptoms of esophagitis and esophageal

obstruction may occur. As the disease progresses there may be difficulty in swallowing liquids, accompanied by retrosternal pressure, loss of weight, dehydration, anemia, and cachexia.

Diagnosis

Esophageal syphilis can be *diagnosed only on the basis of history*, laboratory studies, x-ray findings, esophagoscopy with biopsy and definite rapid response to specific therapy.

Esophagoscopy with biopsy of the lesion is the only positive means of establishing an exact diagnosis of esophageal syphilis. Watson-Williams (39) *states that* "*the local appearances include* (1) general *esophagitis* tending to affect the middle two fourths of the esophagus but in one case the whole, (2) *maceration* of the epithelium with longitudinal desquamation going on to complete shedding of epithelium, (3) formation of *leucoplakic* patches consisting of thickening of the epithelium only."

Treatment

Syphilitic lesions of the esophagus usually respond promptly to antisyphilitic therapy. In some cases however, Levy and Winkelstein (19) report that such therapy, by hastening the healing process with its resulting fibrosis and contraction of scar tissue may not only fail to relieve the obstruction but actually *increase it*. Syphilitic lesions which are causing stenosis should be treated by esophagoscopy and bouginage under direct vision or by bouginage with a previously swallowed thread serving as a guide.

Prognosis

With proper antisyphilitic therapy and bouginage the prognosis of syphilis of the esophagus is good.

DIPHTHERIA

Diphtheria is a specific infectious disease characterized by a local fibrinous exudate or diphtheritic membrane. It is caused by *Corynebacterium diphtheriae*, the Klebs Loffler bacillus.

Incidence

Owing to prophylactic immunization diphtheria is now seldom seen. Its occurrence in the esophagus is exceedingly rare. We have never seen a case of diphtheria involving the esophagus. Reiche (29) recorded esophageal involvement in 11 out of 1000 diphtheria patients studied. Councilman, Mallory and Pearce (7) studied at autopsy 220 cases of diphtheria; they found membrane on the larynx in 75 cases, trachea 66, tonsils 65, epiglottis 60, pharynx 51, nares 43, bronchi 42, soft palate 13, esophagus 12, tongue 9, stomach 5, vagina 2, duodenum 1, vulva 1, ear 1, conjunctiva 1.

Pathology

It is probable that esophageal diphtheria is always secondary to pharyngeal infection. The membrane formation was particularly well studied by Councilman *et al*. Sharply cut undermined erosions were noticeable. The tissue beneath the membrane was intensely hyperemic. The membrane was chiefly fibrinous. Postdiphtheritic esophageal stenosis and paralysis have been noted by Stupka (35), Clerf (6), and Jackson and Jackson (15). Kiviranta (17), however, reviewed the strictures of the esophagus reported as definitely due to diphtheria and concluded that no single case could be positively attributed to diphtheria.

Symptoms

The symptoms may be those of esophagitis or esophageal obstruction; they include substernal pain, dysphagia and regurgitation.

Diagnosis

In the acute stage of diphtheria membranous involvement of the esophagus may be suspected from the history In postdiphtheritic paralysis or esophageal stenosis, x-ray examination should be performed

Esophagoscopy is of value in the study and treatment of post diphtheritic sequelae In paralysis it aids in excluding other diseases of the esophagus In esophageal stenosis esophagoscopy is indicated as an aid to diagnosis, with bouginage for treatment

Treatment

In the acute stage this includes the liberal use of antitoxin and penicillin in addition to the general care of the patient

Prognosis

The prognosis is that of diphtheria in general

ACTINOMYCOSIS

Actinomycosis is a chronic infective disorder produced by *Streptothrix actinomyces*

Incidence

It is a widespread disease occurring nearly three times as often in men as in women The disease is prevalent among cattle and also occurs in the pig We have never seen a case of actinomycosis involving the esophagus

Etiology

Most infections with actinomycosis are probably of endogenous origin since the organism's natural habitat is the human mouth

where it has been cultured repeatedly from gums tonsillar crypts and carious teeth

Pathology

The lesion is like an abscess with large amounts of connective tissue The infection burrows through the tissues for great distances, stopping for nothing and penetrating bones as easily as muscles It involves the digestive tract lungs skin and brain but rarely involves the esophagus In a case reported by Vinson (37) an esophagobronchial fistula occurred The symptoms suggested that the fistula had developed when the patient was 17 years old, or 20 years before she was examined by Vinson

Symptoms

Symptoms referable to the esophagus may include substernal pain dysphagia, regurgitation and sometimes odynphagia Very often there is involvement of the skin of the head and neck from which the organism may be recovered Fever and cough are prominent symptoms in pulmonary actinomycosis

Diagnosis

Because of the organism s saprophytic growth in the mouth and throat discovery of actinomyces in the sputum is not sufficient evidence that the disease in question is caused by the fungus Its gross recognition as sulfur granules and characteristic anaerobic growth which will not proliferate at room temperature makes it easily identifiable (26) X-ray examination of the esophagus may show obstruction but cannot be diagnostic

Esophagoscopy with biopsy is the only means of making a positive diagnosis of esophageal actinomycosis According to Jackson and Jackson (15) the esophagoscopic appearances are those of small ulcerogranulomatous elevations with a tendency to ridge like

formation Jackson and Jackson removed small granules containing characteristic *ray fungi along with granulomatous fragments*

Treatment

In 1948 penicillin was established as the drug of choice in treating actinomycotic infections (25) It should be administered in *large doses for prolonged periods, i e, five million units daily for* two or more months Sulfadiazine is frequently combined with *penicillin Should this combination fail, the broad spectrum antibiotics* and other agents may be utilized Cures have been reported with chlortetracycline (30), chloramphenicol (20), oxytetracycline (18), and isoniazid (11)

Prognosis

The prognosis depends upon the extent of the generalized disease

BLASTOMYCOSIS

Blastomycosis is a fungus disease which usually involves the skin, occasionally involves the lungs and very rarely involves the esophagus *It is caused by a parasitic fungus which grows as a spherical* or oval budding cell

Incidence

Blastomycosis was first described by Gilchrist (10) in *1896* *Since then several hundred cases have been reported* but in only 3 *of them* was the esophagus involved According to Straus (34) *Shepherd and Rhea* (32) and Martin and Smith (23) have each reported a case in which esophageal lesions were demonstrated at post-mortem examination of patients with systemic blastomycosis Vinson Broders and Montgomery (38) reported a case of esophag

eal involvement which was noted clinically and was proved by the use of the esophagoscope together with biopsy, unfortunately however, no autopsy was obtained when the patient died one year later In Shepherd and Rhea s case there were no symptoms referable to the esophagus, but post mortem examination disclosed a small, fluctuating blastomycotic nodule 1 cm in diameter located 7 cm above the cardia In that patient every organ of the body except the brain was involved by the disease We have never seen a case of blastomycosis involving the esophagus

Pathology

The disease is chronic inflammatory infectious and invasive The lesions resemble those of tuberculosis grossly and microscopically

Symptoms

The symptoms may be those of esophagitis or of esophageal obstruction, they may include substernal pain heartburn distress dysphagia, and regurgitation

Diagnosis

The disease must be differentiated from other mycotic infections and from tuberculosis, syphilis and malignant growths Since cutaneous lesions are usually present the diagnosis can be made on the basis of curettings from these lesions *Blastomyces dermatitidis* is identifiable on wet smear as well as by culture The Blastomycin skin test generally indicates past or present infection A complement fixation test also is available (–4) X ray examination may reveal a constricting lesion of the esophagus but cannot provide a specific diagnosis

A definite diagnosis of esophageal blastomycosis can be made only by esophagoscopy and biopsy Jackson and Jackson (15) have reported 4 cases in which the esophagoscopic appearances

were those of "thickening and rigidity of the esophageal wall with granulomas on ulcerated beds in the stenotic lumen" Strauss (34) reported 1 case in which esophagoscopy revealed a soft, granular constricting lesion which was annular and was located 6 cm below the cricopharyngeus muscle

Treatment

Currently 2-hydroxystilbamidine is the therapeutic agent of choice (13 31, 33) The newer anti fungal antibiotics such as rumocidin and mycostatin and the broad spectrum antibiotics are currently being investigated and may be of value The use of iodides is empirical and may even affect the disease adversely

Stenotic lesions of the esophagus should be treated by dilatation under direct vision through the esophagoscope

Prognosis

The prognosis of cutaneous blastomycosis is good as regards life expectancy, but its chronic course may extend many years Complete excision may result in cure In systemic cases the prognosis is poor The combination of complement fixation test and cutaneous test has been reported to be of value in prognostication A favorable outcome may be expected with a positive skin test and a negative complement-fixation reaction A high titer of complement fixing antibodies and a negative skin test offers a poor prognosis

Pulmonary tuberculosis was found to be present in Vinson s case It was also found in one of Jackson s cases in which there were lesions in the upper-lobe bronchi as well as in the esophagus

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never any proof that the stenosis was the result of the pellagra, the two diseases may have been merely coincidental



FIGURE 103 Benign esophageal stenosis Rather high obstruction seen in a pellagra patient before bouginage

In each of Fisher's 17 cases a 9 mm esophagoscope was passed under local anesthesia All examinations revealed an esophageal mucosa which was intensely hyperemic and in some cases was edematous Multiple tiny ulcerations of the esophageal mucosa were noted in 9 cases No tumor or foreign body was found in any case In our own case the lumen appeared red, granular, and narrowed sometimes with sufficient irregularity and ulceration to suggest neoplasm, however, none of the eight biopsy specimens obtained at different times ever showed malignancy On some occasions there was definite sloughing of the mucosa

TREATMENT

Nicotinamide should be given in large doses — for an adult, 100 mg by mouth 3 times a day with meals Up to 300 mg by mouth

may be given with safety 3 times a day In a very sick patient, or one with esophageal obstruction nicotinamide may be given intramuscularly or intravenously, 50 mg 3 times a day For protein deficiency, brewers' yeast should be given in doses of 1 to 3 tablespoonfuls by mouth 3 times a day



FIGURE 104 Close up view of the same stenotic area after bouginage

Each patient in Fisher's series was given 150 mg of nicotinamide, 15 mg of thiamin chloride and 15 mg of riboflavin daily for 3 months After this they were given normal daily maintenance doses of vitamins considered essential for health Our own patient was treated intensively for pellagra but in spite of everything including many transfusions over a three-year period she succumbed to pellagra hypoproteinemia nutritional anemia and acute pulmonary edema the final episode occurring at the age of 45 with an acute anaphylactic reaction of unknown cause

Prognosis

Judging from our case, the prognosis may be bad, but in the series studied by Fisher all of the patients were able to swallow solid food without experiencing dysphagia after three weeks to two months All of his patients *improved tremendously in their general* health and the dermatitis and stomatitis continued to show improvement in all cases

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Scleroderma

29

Scleroderma is a condition of localized or diffuse induration of the skin. The basic process is sclerosis of the connective tissues of the body. According to O'Leary (5) there are four dermatoscleroses: scleroderma, acrosclerosis, scleredema and dermatomyositis, in all of which a certain degree of cutaneous sclerosis is part of the clinical picture.

Incidence

Ehrmann (2) in 1903 was the first to describe progressive dysphagia and esophageal disturbances in scleroderma. In 1945 Olsen, O'Leary, and Kirklin (6) reported esophageal lesions associated with acrosclerosis and scleroderma. They believe that acrosclerosis differs from scleroderma in that the trophic disorder in the former appears to have its origin in vasomotor disturbances. Of 32 cases reported, 25 patients were female and 7 male. Twenty-four cases showed either stenosis in the lower esophagus, lack of motility, dilatation, or cardiospasm. Scleroderma of the esophagus is usually a disease of middle life. Spontaneous recovery has been observed in children.

Pathology

Grossly there is sclerosis of the connective tissue of the esophagus, the organ is usually found to be dilated and to have a thickened mucosa which may have a varnished appearance Leukoplakia and ulceration may be present Weissenbach, Steward, and Hoesli (8) report that histologic studies show that the mucosa is apparently not involved The submucosa is indurated and thickened and is arranged in thin layers Often there is moderate round-cell infiltration of the mononuclear type, sometimes rare polymorphonuclear leukocytes are seen The elastic tissue may be very dense but more frequently appears degenerative The muscularis mucosa is hypertrophied more rarely atrophied The tunica muscularis occasionally hypertrophied, is atrophied in most cases Often there is a rich vascularization with dilated vessels distended with blood, the arterioles are almost always hypertrophied There is no abnormality of the ganglia in the nerve plexi as observed in achalasia the dysphagia being produced by the atrophy and fibrosis of the esophageal musculature

Symptoms

Esophageal symptoms seldom appear early in the disease They usually occur later, when other symptoms are already established According to Weissenbach *et al* (8) it is in the cases with marked atrophy of the mucosa that dysphagia presents itself with a particularly characteristic symptom complex It is usually two or three years after the onset of the illness or sometimes even later that swallowing difficulties appear In the late stages of the disease difficulty in swallowing becomes more marked it becomes impossible to swallow solid food Sometimes only a gastrostomy will save the patient On the other hand there are many cases in which there are no severe esophageal troubles despite accentuation of the cutaneous sclerosis

Hale and Schatzki (3) examined by x ray the upper gastrointestinal tract of .. patients (15 females and 7 males) who were

known to have scleroderma varying in duration from five months to 22 years In 7 of the group examination of the esophagus was made because of dysphagia The remaining 15 had no definite symptoms that were referable to the esophagus, these were examined routinely for the purpose of this study In 6 of the group with no symptoms however careful questioning disclosed that a sensation of material remaining in the mid chest and occasionally localized burning or pain after eating hurriedly or after eating a large meal, had been noticed In the cases reported by Lindsay Templeton, and Rothman (4) dysphagia and burning substernal pain were the chief symptoms

Diagnosis

The diagnosis of scleroderma is made chiefly by physical examination which shows scleroderma of the skin and connective tissue When there is a history of dysphagia or substernal pain esophageal involvement should be suspected X ray examination (3 4) is of great importance and conforms to a certain pattern as follows the time of transit of the barium from the pharynx to the stomach is prolonged, in some cases complete emptying of the esophagus is delayed for as much as half an hour Peristalsis of the esophagus is decreased (Figs 105 106)

Dornhorst Pierce and Whimster (1) studied 6 consecutive patients with scleroderma by x rays and esophageal contractions Although none of these patients complained of dysphagia all had a definite abnormality of esophageal function The characteristic defect was failure of the propulsive waves to continue into the lower half of the esophagus the upper half contracted with normal vigor The weakness of the lower esophagus was manifested radiographically when the patient was observed swallowing in the Trendelenburg position The bolus was carried briskly to about the position of the tracheal bifurcation where its progress ceased These workers concluded that the essential disorder was weakness and wasting of the muscles of the lower esophagus

Esophagoscopy is often very difficult in patients with scleroderma. Because of a cutaneous tightness in the neck or about the mouth, these patients are frequently unable to extend the head or open the mouth adequately, hence general anesthesia may be re-



FIGURE 105. X-ray appearance of scleroderma of the esophagus, showing considerable barium retention after an interval of 15 or 20 minutes. Very little peristaltic activity is seen.

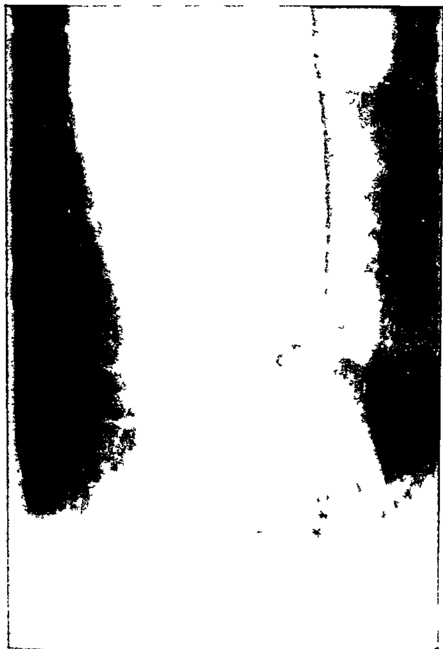


FIGURE 106 X ray appearance of scleroderma of the esophagus in a 50-year-old woman (Courtesy of Dr Richard Schatzki) The x ray shows constriction at the junction of the esophagus and the herniated stomach Emptying time of the esophagus is delayed Esophagoscopy revealed red edematous mucosa with a lumen only about 4 mm in diameter at the level of the upper end of the hernia The patient improved after bouginage

quired Esophagitis with exudation, erosion, ulceration and stricture formation may be found Biopsy may show inflammatory changes absence of the epithelial layer, thickening of the mucosa and connective-tissue changes in the submucosa that suggest scleroderma

TREATMENT

The treatment of esophageal scleroderma is dilatation of any strictures which may be present This can be carried out at the time of esophagoscopy or by means of bougies passed with a previously swallowed thread as a guide If bouginage is unsuccessful surgery should be considered Cortisone has been found very helpful in the treatment of generalized scleroderma (7)

Surgery becomes necessary when the fibrosis of the disease results in narrowing and obstruction that are not amenable to bouginage

Because of the generalized nature of the disease these patients may not tolerate surgery well, and they should be prepared with care *It is possible that corticosteroids may be useful adjuvants* Fortunately the areas of narrowing are frequently small and well *localized*, hence corrective surgery may usually *be limited to some* of the plastic procedures detailed in Chapter *11* for benign peptic stenosis

The surgeon must remember that the sclerodermatous patient remains susceptible to other causes of esophageal inflammation and stenosis If a hiatus hernia is present at the time of surgery, it also should be corrected Shortening of the esophagus due to scleroderma *per se* may produce a sliding type of hiatus hernia

PROGNOSIS

The prognosis should be guarded

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Myasthenia Gravis

30

Myasthenia gravis is a chronic, progressive muscular weakness commonly affecting the eyes and facial musculature, producing dysarthria and dysphagia, or presenting a weakness that either is general or is localized to the arms or legs. It is not accompanied by atrophy.

Pathology and Etiology

Jolly (6) *first focused attention on the pathologic process in myasthenia gravis with his statement that it appeared to be a defect in the neuromuscular junction with a consequent inability of the nerve impulse to exercise its effect upon the effector organ.* The etiology of this defect of neuromuscular transmission is still under investigation. It is believed to be due to some disturbance *in* the metabolism of acetylcholine. Weigert (15) *first noted the presence* of a thymic tumor in a case of myasthenia. The resemblance of the disease to curare poisoning stimulated Walker (14) to use physostigmine and prostigmine — with great success.

Keynes (7) found 41 tumors of the thymus among [illegible]19 patients, an incidence of 15.4 per cent. Reviewing 47[illegible] cases of myasthenia at the Mayo Clinic, Eaton and Clagett (5) found 77 tumors (16 per

cent) Castleman and Norris (4) found thymoma… bout 25 per cent of 330 cases collected from the literature B… …leman (3) and Seybold *et al* (10) have described patients wh… …d thymomas but no evidence of myasthenia There are no fe tures in the histology of thymomas which would …pa ate he case with myasthenia from those without sympto… Thymic tumors may be benign or malignant, when they ar… …lignant they spread by local invasion, not by metastasis

INCIDEN…

Patients who have myasthenia but n… …m i show a predominance of females, whereas the sex distribu… …is about equal among those who have thymoma The incidence of tumors increases with age Myasthenia without thymoma is considered a disease of the relatively young (3)

SYMPTOMS

The condition was first described by Thomas Willis (16) in 1672 The following excerpt from his article is descriptive of the typical patient

" Wherefore, when the spirits inhabiting the Brain are conscious of the debility of others disposed in the members they themselves refuse local motions for that it would be too difficult a task to impose on their companions wherefore the sick are scarce brought by any persuasion to try whether they can go or not nevertheless those laboring with a want of spirits who will exercise local motions as well as they can in the morning are able to walk firmly, to fling about their arms hither and thither or to take up any heavy things, before noon the stock of the Spirits being spent which had flowed into the muscle they are scarce able to move hand or foot At this time I have under my charge a prudent and an honest woman who for many years hath been obnoxious to this sort of spurious Palsy not only in her members but also in her tongue, she can for some time speak freely and

readily enough, but after she has spoke long, or hastily, or eagerly, she is not able to speak a word but becomes as mute as fish, nor can she recover the use of her voice under an hour or two "

According to Viets (13), dysphagia is the first symptom in about 20 per cent of patients Thirty-five of the 175 patients he studied had dysphagia or dysarthria (nasal speech) or both In addition dysphagia may occur in the course of the disease, at which time it is usually indicative of an exacerbation In Blalock's (1) series of 20 patients treated by thymectomy for myasthenia gravis, 14 complained of difficulty in swallowing

Diagnosis

Diagnosis is confirmed by the neostigmine test After the amount of weakness is ascertained, the patient is given 1 5 mg of neostigmine and 0 6 mg of atropine Disappearance of the weakness within thirty to sixty minutes with return of symptoms after four to eight hours is considered a positive test For further details the reader should consult the article by Schwab and Viets (9) According to Viets, no disease except myasthenia gravis gives rise to such a complete return of normal function of the swallowing reflex after the appropriate amount of neostigmine is given and the patient is allowed to absorb the medicine over a period of twenty to thirty minutes

X-ray examination (Fig 107) usually shows that the barium is retained in the pyriform sinuses and has perhaps spilled over into the larynx and trachea

Esophagoscopy is indicated only when some other disease is suspected as the cause of the dysphagia Except for the fact that the usual muscle tone of the cricopharyngeus is absent esophagoscopy is negative in myasthenia gravis

Treatment

Treatment of patients with myasthenia is best carried out by a qualified neurologist and internist using neostigmine and other



FIGURE 107 X ray appearance of myasthenia gravis. Barium sticks in the pharynx and flows into the nasopharynx After the administration of physostigmine barium flows freely through the pharynx.

related compounds Because of the variable nature of the disease and the frequency of spontaneous remission, results of treatment have been difficult to evaluate

Because of the possible relation between myasthenia and thymic

hyperplasia or enlargement thymectomy has been advocated as a *method of treatment* Blalock (1, 2) was the first to remove a thymoma with alleviation of the myasthenia symptoms The results of thymectomy have, however, been equivocal Schwab and Leland (8) found that all patients with thymomas and most males with non-neoplastic thymus glands derived little benefit from operation The best results were found in females under the age of 30, this group had a remission rate of 68 per cent as compared with a control figure of 34 per cent Eaton and Clagett (5) compared 62 myasthenia patients who underwent thymectomy with 56 who had not been treated surgically They found a 35 5 per cent *remission after surgery as contrasted to 28 5 per cent spontaneous* remission in the second group They did not consider that thymectomy alleviated the condition Keynes (7) has advocated preoperative x-ray therapy in all thymomas to improve the poor results in this group

Despite the poor results in the thymoma group, surgery is always indicated for removal of the tumor if not for treatment of *the myasthenia, which may or may not be present*

At the present time it would seem advisable to limit surgery to selected patients and to those who have not satisfactorily responded to drug therapy and in whom the chance for spontaneous remission is small Pregnancy has resulted in complete remission of symptoms and myasthenia has developed in patients after removal of a thymoma

Patients with myasthenia are often very *poor surgical risks* Thymectomy should not be performed as an emergency procedure, the patient should be well regulated and stabilized before operation Neostigmine should be given by intravenous drip during operation and a qualified neurologist and anesthetist should be present

Irradiation of the thymus gland for this disease has few advocates however, it may be *useful in the preparation of a poor-*risk patient for surgery and as a preoperative measure in the thymoma group

ACTH has been reported to produce remission, but there is some doubt of this (11, 12)

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Paralytic Dysphagia

31

Paralysis of the muscles of deglutition is a central nervous system disease of bulbar origin

Etiology

According to Kubik (4), the various causes of paralytic dysphagia can be listed in the order of frequency as follows

(1) Poliomyelitis — the bulbo-spinal type (2)
(2) Progressive bulbarpalsy
(3) Infarction of the medulla
(4) Diphtheritic neuritis
(5) Infectious polyneuritis with paralysis of the tenth nerve
(6) Nerve trauma with glossopharyngeal paralysis

Naffziger Davis and Bell (6) have reported a bullet wound injury to the ninth, tenth eleventh, and twelfth cranial nerves with inability to swallow they describe in detail an ingenious surgical repair by muscle transplantation Other rare causes include abscess of the medulla tuberculoma of the medulla and meningovascular syphilis Dysphagia is also very marked in rabies in which disease the patient dreads even the sight of water (hydrophobia) because of the resultant exceedingly painful spasms of mouth

larynx, and pharynx. Another rare cause of dysphagia is botulism, which may cause paralysis of any of the motor cranial nerves.

Adams (1) recently discussed a patient whose *major complaint* was dysphagia. In this patient there was a defect in initiating the *swallowing movement. According to Adams, "one encounters* disturbances in the swallowing mechanism not infrequently. *Patients with pseudobulbar palsy — that is, with lesions in the* corticobulbar tracts bilaterally — lose the capacity to initiate the act of swallowing and have dysarthria and dysphonia as well. The tongue retains its bulk, and the jaw reflex is exaggerated, in contrast to bulbar palsy (poliomyelitis, diphtheria, progressive bulbar palsy), in which the tongue becomes atrophic. Then, too, a curious difficulty in swallowing is frequently seen with unilateral *lesions in the medulla. In the Wallenberg syndrome, usually produced* by a lateral medullary infarction, the patient may lose the capacity to swallow *for weeks or months, others, however, if the* lesion is small have only the slightest trace of dysphagia. In addition, coughing, hawking and clearing the pharynx of secretions are impaired. Thus a unilateral affection of the vagus complex will certainly disturb the act of swallowing."

Other syndromes (associated with laryngeal paralysis) which may cause dysphagia are those of Vernet and Avellis (3, 5). In the *former there is paralysis of the larynx, pharynx,* soft palate and sternocleidomastoid and trapezius muscles. In the latter there is *paralysis of the larynx and soft palate on the same side* some involvement of the pharyngeal constrictors and sometimes contralateral loss of pain and temperature sensitivity in the neck, trunk and extremities. We were recently quite confused in establishing the diagnosis of Avellis's syndrome in a patient who had had excision of carcinoma of the tongue with right hemimandibulectomy and right radical neck dissection. Nine months later he developed dysphagia and when an esophagram suggested a lobulated filling defect in the distal hypopharynx at the junction of the cervical esophagus, recurrent carcinoma was suspected. Esophagoscopy, however, was negative and laryngoscopy showed the right vocal cord moved

poorly Further x ray study established the diagnosis of the syndrome of Avellis, which in this case was probably due to thrombosis of the posterior cerebral artery involving the tenth and eleventh nerves Although most patients with this syndrome recover in a few months, further thrombosis occurred with fatal termination about one month after the diagnosis was established



FIGURE 108 X ray appearance of paralysis of pharynx. Note barium retention in valleculae and in the pyriform sinus.

SYMPTOMS

When the muscles of deglutition are completely paralyzed the patient is unable to swallow either liquids or solids Overflow of *secretions or food into the larynx* often causes spells of choking or coughing

DIAGNOSIS

The diagnosis may be easy if paralysis is thought of as a cause of dysphagia The history, physical examination, and laboratory findings should suggest the diagnoses enumerated above X-ray examination usually shows that the barium is retained in the pyriform sinuses (Fig 108) and may be spilled into the larynx and trachea

Esophagoscopy *is not indicated in ordinary cases of paralysis* Where the diagnosis is in doubt, however, or where foreign body is suspected, esophagoscopy should be performed to exclude *mechanical obstruction* or *to remove the foreign body*

TREATMENT

The cause of the paralysis should be ascertained and suitable treatment instituted Feeding *tubes can readily* be passed *through* the nose or mouth in order to maintain nutrition and fluid balance Special care must be taken to avoid aspiration pneumonitis

PROGNOSIS

The prognosis is that of the disease which has caused the paralysis

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Emotional Disorders of Swallowing

32

All disorders of swallowing for which no organic cause can be found may be included under the heading of "emotional disorders of swallowing." Various other terms have been applied, including "globus hystericus," "hysterical dysphagia," "functional" or "psychosomatic dysphagia," and "spasm of the esophagus not associated with any demonstrable organic disease."

Incidence

Emotional dysphagia is exceedingly rare. Dejerine (1) in 1911 described 6 cases of hysterical dysphagia in his monograph on psychoneuroses. Hurst (2) knows of no other recorded cases of hysterical dysphagia and says that in his own experience it is remarkably rare. Whereas in the First World War he saw over 100 cases of hysterical aphonia and over 50 of hysterical vomiting in soldiers, he saw no case of hysterical dysphagia. Many years ago Hurst made such a diagnosis in a few middle aged women, but he is now convinced that the majority were examples of the upper

dysphagia-and-anemia syndrome of Paterson (3) which is easily mistaken for hysteria as it was by Plummer and Vinson (4) We agree with Hurst that there is nothing hysterical about the dysphagia-and-anemia syndrome (see Chapter 13)

Etiology

The causes of emotional dysphagia may sometimes be traced to cancerphobia Some friend or relative of the patient may be suffering from cancer of the esophagus and the patient may then imagine that he himself has difficulty in swallowing Sometimes the emotional disturbance begins when a patient has difficulty in swallowing a pill or he may be frightened into a state of emotional dysphagia by the belief that he has accidentally swallowed some poisonous substance

Symptoms

Some patients with emotional dysphagia will not even attempt to swallow The patient reported by Hurst continued to chew and did not appear to make any effort to swallow although she spoke of the food sticking in her throat Other patients however can swallow food but experience difficulty in doing so and the food may quickly be regurgitated in whole or in part

Diagnosis

All organic diseases of the esophagus must be excluded before a diagnosis of emotional dysphagia is made Even when an emotionally unstable person presents himself with a somewhat atypical history for organic disease one should not make a diagnosis of emotional dysphagia without a careful clinical study including x-ray examination and esophagoscopy It should never be forgotten that emotionally unstable persons often do have organic disease Malingering should be considered in the differential diagnosis

Esophagoscopy is negative in emotional dysphagia The performance of esophagoscopy in this condition is usually indicated to exclude organic disease, and in reassuring the patient it may be strikingly beneficial

Treatment

Dysphagia due to emotional causes requires careful handling on the part of the physician Since the patient regards his trouble as a very serious one as indeed it is, the physician must not dismiss it lightly The patient should never be told that there is nothing the matter with him After a negative x ray examination and negative esophagoscopy, it is usually wise to explain to the patient that no organic disease is demonstrable and that since a metal tube has readily reached the stomach food also will pass normally Such an approach to the problem may result in complete cure If symptoms persist, however it may be advisable to consult a psychiatrist who should make a complete study of the patient's emotional background and make every effort to eliminate environmental strains It is very important that the patient have complete confidence in all the physicians who are handling his case

Prognosis

If organic disease has been completely excluded and the patient is carefully treated as outlined above, the prognosis is excellent with complete restoration of the swallowing function

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